MANUAL OF

Rheumatic Diseases

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The YEAR BOOK PUBLISHERS, *Inc*

200 EAST ILLINOIS STREET, CHICAGO

PRINTED IN U S A.

This book is respectfully dedicated to the general practitioner, for it is he who first sees these patients with chronic arthritis and has the opportunity to prevent crippling and deformity This illustrated manual has been prepared in the belief that it may provide hope and better care for the seven million victims of chronic rheumatism in the United States.

Preface



IN THIS BOOK an attempt has been made to simplify and condense the practical information on rheumatic diseases so that it can be used quickly and easily as a guide to diagnosis and treatment by any interested physician Large laboratories and well equipped hospitals are not required to carry out most of the simple basic routine described here Procedures of office and home practice are emphasized No effort has been made to write a documented scientific treatise It is not intended to cover all aspects of all rheumatic diseases nor to discuss in detail the rare forms. Theories have largely been omitted. The reader seeking more detail or references to original articles should consult larger texts on the subject The Rheumatism Review prepared by the American Rheumatism Association usually appears in the *Annals of Internal Medicine* every two years It is an excellent current reference

We are indebted to Mrs. Edna McCarthy, physical therapist, for her assistance in developing useful prevention and correction technics and to H D Moore and F O. White for providing much of the photographic material used in Chapter 9

—W.P H
—D.F.H.

Table of Contents

The Problem of Arthritis and Rheumatism

THE TERM "rheumatism" is as old as medicine and has been used for centuries to designate all forms of painful skeletal disturbances, whether joints, muscles, fibrous tissue or nerves were involved "Rheumatism" is still used and is perhaps the best family name to include all rheumatic diseases Current preference, however, is for the expression "arthritis and rheumatism," which simply means arthritis and the other rheumatic diseases.

Importance —It is estimated that in the United States approximately seven and one-half million people are suffering from some form of chronic rheumatic disease. Therefore, approximately 1 in each 20 of our population is affected If the average family consists of four individuals, then more than thirty million people have one member of their family suffering from chronic rheumatism. According to public health figures, there are twice the number of patients with chronic rheumatism as there are with heart disease, 10 times as many as have tuberculosis, 10 times as many as have diabetes, 7 times as many as have all forms of cancers and

tumors and 40 times as many as have infantile paralysis Arthritis and rheumatism cause more years of disability and crippling than all accidents combined.

Because arthritis only makes cripples for a lifetime but does not kill, it has failed until recently to stir the interest of the physician and the public sufficiently to do anything about the problem In 1947, approximately $20,000,000 was collected by public subscription for infantile paralysis, about $18,000,000 for tuberculosis and several millions each for cancer and heart disease. These are worthy causes, but in that year not one cent was raised by public subscription for arthritis and rheumatism One seldom read anything in magazines or newspapers or heard mention over the radio of rheumatism or arthritis

In the spring of 1948, the American Rheumatism Association, a group of physicians interested in these diseases, and interested laymen formed the Arthritis and Rheumatism Foundation, a nonprofit organization committed to raise money for research into the cause and cure of these diseases, to provide professional and public education and to do everything possible to improve the knowledge and treatment of these crippling diseases. The Arthritis and Rheumatism Foundation's activities, together with recent advances in research on rheumatic diseases, have created an increased public awareness of the problem. With physician and lay support, the hope for an early solution of many of these ~roblems is bright.

Since the newspaper and magazine reports of the miracles chieved with cortisone and ACTH, patients and perhaps me physicians are apt to neglect the necessary routine rocedures and wait until the miracle drugs are available.

The answers to this line of thinking are simple and blunt.

1. No amount of magic medicine, if it were available, would restore destroyed joints nor correct deformities.

2. Enough cortisone and ACTH cannot be obtained from present sources to be available for general use, even if the price were low enough

3 Next to nothing is known about long-time administration or the toxic effects that may be produced by administration of these drugs.

4 Nothing is yet known regarding the mechanism of action in relieving rheumatic diseases.

5 Synthetic products offer hope, but not immediately

6. The brightest hope lies in the remarkable and dramatic value of cortisone and ACTH as research tools It is possible with these substances to produce remissions in the disease at will, and when sufficient research work has been done the mechanism of the remission will undoubtedly be revealed Unfortunately, that may not come this year or next.

7. Each day that crippling arthritis exists, destruction of joints and deformity is slowly progressing It is therefore essential *now* to do everything possible to prevent deformity, to maintain joint motion and to do the many helpful things that can be done for the patient if he is to benefit ultimately from the specific cure when it is available

CLASSIFICATION

This classification of diseases of joints and related structures is taken from the one adopted by the American Rheumatism Association

1 Rheumatoid arthritis
   a) of peripheral joints

b) of spine—rheumatoid spondylitis

2. Degenerative joint disease (osteoarthritis, hypertrophic arthritis)

a) localized

b) generalized

3 Fibrositis

a) localized

b) generalized

4. Arthritis due to gout

5 Arthritis due to specific infections, e g., gonococcic, tuberculous, pneumococcic, syphilitic, brucellar, typhoid, meningococcic, suppurative, Haverhill (rat bite) fever, dysenteric, mycotic (coccidioidomycosis) and others

6. Arthritis due to rheumatic fever

7. Arthritis due to trauma

8 Neurogenic arthropathy

9 New growths

10 Hydrarthrosis, *intermittent*

11. Miscellaneous diseases, e.g., disseminated lupus erythematosus, polyarteritis, scleroderma, drug intoxication, serum sickness, hemophilia, purpura, acromegaly and hysteria.

This classification may seem quite cumbersome and confusing, but actually approximately 95 per cent of all chronic rheumatism is covered by the first four headings, i e., rheumatoid arthritis, degenerative joint disease, fibrositis and gout, and it is these four that will receive chief attention in the following pages.

CHAPTER 2

Rheumatoid Arthritis

OF ALL THE RHEUMATIC DISEASES, rheumatoid arthritis is perhaps the greatest crippler and our most difficult therapeutic problem. It is a constitutional disease which manifests itself as a profound systemic disturbance and by chronic, deforming, progressive polyarthritis There is no known single cause and no single specific cure

DIAGNOSIS

Once the disease is well established the diagnosis is usually not difficult. The characteristic picture of chronic polyarthritis, with soft tissue swelling, pain and tenderness in the proximal interphalangeal and metacarpophalangeal joints, the wrists, elbows, knees, ankles and metatarsophalangeal joints, or any combination of these joints, is almost diagnostic. These changes are usually associated with weakness, muscle atrophy, evidences of constitutional illness, flexion deformities, rapid sedimentation rate, demineralization of adjacent bone and cartilage loss The onset of rheumatoid arthritis may be insidious or as acute as that of rheumatic fever Diagnostic difficulties usually present them-

selves in the early or minimal stage of the disease process.

Diagnostic points in rheumatoid arthritis (peripheral)—
1 In 95 per cent of individuals who have rheumatoid arthritis it develops after the age of 16 (approximately 5 per cent occurs in children).

2. Onset of rheumatoid arthritis is insidious in approximately 90 per cent and acute or subacute in approximately 10 per cent.

3 Proximal interphalangeal, metacarpophalangeal, wrists, metatarsophalangeal, knees, elbows and ankles are the joints most likely to be involved and in that approximate order of frequency (Figs. 1–12). However, any joint may be involved.

4. Swelling around the joints is soft and there is usually marked tenderness to firm pressure over the joint.

5. More than one joint is usually affected, and involvement is often symmetrical. Single joint involvement is seen occasionally and occurs in atypical rheumatoid arthritis or in the very slow, insidious form of the disease.

6 Usual constitutional symptoms are fatigue, loss of appetite, low grade fever, loss of weight, nervousness and depression.

7. Weakness is out of all proportion to pain, joint disease or muscle atrophy Test grip, using two of the examiner's fingers, is often conspicuously reduced even early in the disease (Fig. 13, p. 22).

8 Circulatory disturbances are frequently noted in the form of cold clammy hands and feet and "liver palms."

9. *Painful stiffness* is present especially after rest and is *an almost constant feature of the disease It is more painful* and its character is different from the stiffness occurring in fibrositis and osteoarthritis.

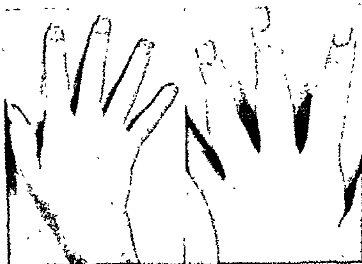


Fig. 1 *(top left)* —Early rheumatoid arthritis. Note metacarpophalangeal swelling

Fig. 2 *(top right)* —Early fusiform changes in proximal interphalangeal joints

Fig. 3 *(bottom)* More advanced stage, with wrists, metacarpophalangeal and proximal interphalangeal joints involved and beginning deformity



FIG. 4 *(top)*.—Rheumatoid arthritis, with characteristic wrist flexion and ulnar finger deviation.

FIG. 5 *(bottom left)*.—Knee involvement, with swelling and atrophy.

FIG. 6 *(bottom right)*.—Same case as preceding, showing pronounced atrophy of the quadriceps muscle.





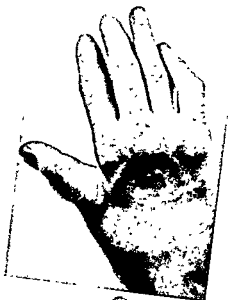
FIG. 7 *(top)* —Rheumatoid arthritis of only a few months' duration, but with muscle atrophy already developed.

FIG. 8 *(bottom)* —Typical ankle and tarsal involvement.



Fig 9 *(top)* —Rheumatoid arthritis with conspicuous thickening of the wrist

Fig. 10 *(bottom)* —Advanced arthritic deformity, subluxation and disarticulation of the fingers



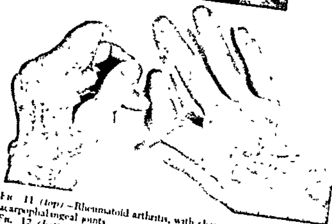


Fig. 11 (*top*).—Rheumatoid arthritis, with characteristic deformity of the metacarpophalangeal joints.

Fig. 12 (*bottom*).—Long-standing disease, with atrophy of the interosseous muscles.

10 Muscle atrophy is always present quite early and increases with activity and duration of the disease. It is seen well in the interossei (Fig. 12) and quadriceps muscles (Fig. 6).

11. X-rays made late in the disease are diagnostic but are seldom helpful in early stages. Figures 14–28 show the changes from normal to early disease as compared with advanced changes. Absence of positive x-ray evidence *does not* rule out early rheumatoid arthritis. Subcortical cysts or areas having a punched-out appearance are common in rheumatoid arthritis, they are often misdiagnosed as gout (Figs 29 and 30, p. 29).



FIG 13 —Test grip, a valuable indicator in the early stages of rheumatoid arthritis. Here there is no deformity (the disease is in an early stage), but grip is weak

12 Anemia is nearly always present once the disease is well established. The anemia is microcytic and hypochromic (low color index). Although it resembles an iron deficiency

anemia, the response to iron is not ordinarily satisfactory.

13. The erythrocyte sedimentation rate is almost always increased if the disease is established and active. Rarely, one sees a patient with active rheumatoid arthritis who has a normal sedimentation rate. It is well to ascertain which method is being used to determine the sedimentation rate because the normal value varies with each method.

14. Spontaneous remissions and exacerbations are said by some workers to be the natural course of the disease This statement is misleading It is perfectly true that patients tend to have cycles of being better and worse, but once rheumatoid arthritis is definitely established, complete, 100 per cent spontaneous remissions are extremely rare Continuous chronic involvement of joints since the onset of the disease, without a single period in which there was 100 per cent recovery, is a strong point in favor of the diagnosis of rheumatoid arthritis.

15 Loss of motion, particularly of the wrists, and loss of full flexion of the fingers may suggest early rheumatoid arthritis

Differential diagnosis —The following conditions may be confused with and should be distinguished from rheumatoid arthritis

1 RHEUMATIC FEVER —Typical rheumatic fever is a short-term disease, without joint sequelae Usually in rheumatic fever the carditis, the migratory joint disturbances, the history of a respiratory infection and dramatic response to salicylates make the diagnosis relatively easy. Occasionally, rheumatoid arthritis of acute onset cannot immediately be differentiated with certainty from rheumatic fever Close observation, however, for a week or two will almost certainly

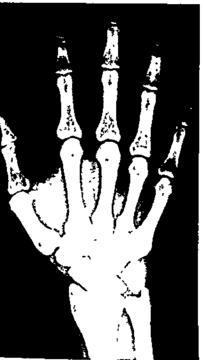


Fig. 14 *(left)*.—Normal hand

Fig. 15 *(center and right)*.—Early rheumatoid arthritis, showing juxta-articular demineralization

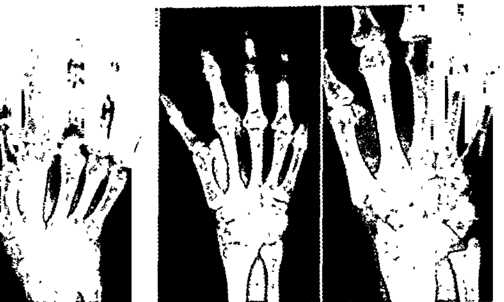


FIG. 16 (*left*) — Rheumatoid arthritis definitely established. Cartilage destruction is most apparent in the carpals.

FIG. 17 (*center*) — Advanced stage, with subluxation of the metacarpophalangeal joints and almost complete loss of identity of the carpals.

FIG. 18 (*right*) — Advanced wrist changes and typical cystic areas in the carpals and heads of the metacarpals.

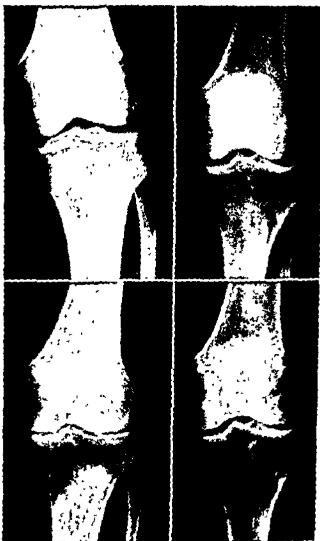


Fig 19 *(top left)* —Normal knee, anteroposterior projection.
Figs 20–22.—Early rheumatoid arthritis, showing gradual increase i demineralization.



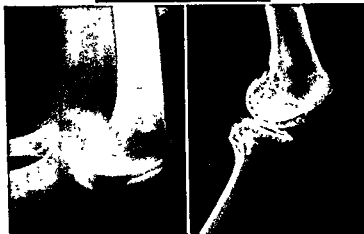


Fig. 23 *(top)*—Normal knee, lateral projection.
Figs. 24 and 25 *(bottom)*—Early rheumatoid arthritis, showing demineralization.

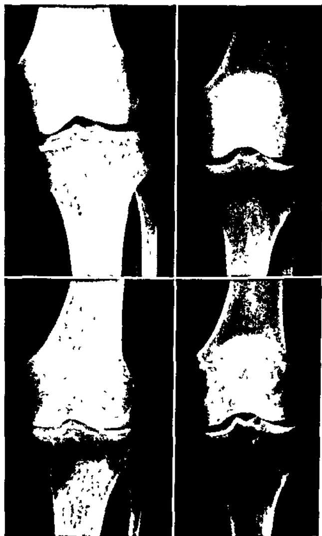


Fig. 19 *(top left)*—Normal knee, anteroposterior projection.
Figs. 20–22.—Early rheumatoid arthritis, showing gradual increase in demineralization.





FIG. 23 *(top)*—Normal knee, lateral projection
FIGS 24 AND 25 *(bottom)*—Early rheumatoid arthritis, showing demineralization



Figs 26–28.—Advanced rheumatoid arthritis, showing obliteration of the joint space, deformity of articulating surfaces and secondary hypertrophic changes

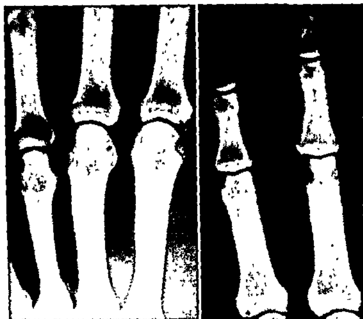


FIG 29 *(left)*.—Early rheumatoid arthritis, with typical small subcortical cysts

FIG 30 *(right)*.—Punched-out areas near the proximal interphalangeal joints, frequently seen in rheumatoid arthritis and often erroneously attributed to gout

settle the diagnosis. In acute rheumatoid arthritis, fever may be as prominent as in rheumatic fever.

2 ATYPICAL RHEUMATOID ARTHRITIS.—*a)* "Palindromic rheumatism" is mentioned here not to establish points of differentiation from rheumatoid arthritis but to emphasize that it is an atypical form of rheumatoid arthritis. Palindromic rheumatism is characterized by sudden acute attacks of pain and swelling, usually in a single joint, often lasting no longer than 24 hours and ending in complete recovery. These attacks may continue for years without showing any tendency to become chronic, but as a rule the attacks gradually become more frequent until recovery is not complete between the attacks and the disease progresses to chronic arthritis which behaves typically as rheumatoid arthritis. In short, it may be said that palindromic rheumatism is rheumatoid arthritis with a life history much like gout. We have watched a number of such palindromic manifestations progress to typical rheumatoid arthritis. The disease responds to the therapy used for rheumatoid arthritis.

b) Intermittent hydrarthrosis. This condition is seen most commonly in the knees, where one or both may become quite swollen without development of any very striking constitutional symptoms and little or no severe pain. These swellings may come and go once or more times a year, with complete recovery between attacks. Again, as in palindromic rheumatism, the swelling usually becomes chronic and may even persist in a chronic form for several years before other joints are affected or the full symptoms of rheumatoid arthritis appear In these patients, if watched for a long enough time, typical rheumatoid arthritis will usually develop, and it is our conviction that this is another form of atypical

rheumatoid arthritis. Therapeutic procedures are identical with those for rheumatoid arthritis.

c) There are other bizarre and atypical forms, for rheumatoid arthritis is a constitutional disease, but the diagnosis can usually be made if it is included in the differential diagnosis

3 PSORIATIC ARTHRITIS.—Authorities disagree as to whether there is a definite, specific syndrome of psoriatic arthritis or whether there is simply an association of the two diseases. Certainly one sees many patients with rheumatoid arthritis who also have psoriasis. We are convinced that when psoriasis develops around a finger- or toenail, with arthritic involvement of the proximal joint and beyond, we are justified in making a diagnosis of psoriatic arthritis. Figures 31 and 32 illustrate nail and joint involvement. In such cases, clearing of the psoriasis often produces dramatic relief from the arthritis. It is only fair to say, however, that even without this particular type of involvement—when the body is well covered with psoriasis—much improvement of the arthritis often follows the clearing of the psoriasis. It is of no great importance whether one wishes to make the distinction of this type of arthritis or not, as the treatment of the joint problem is identical with that of rheumatoid arthritis. It is certainly true that, in addition to caring for the joints, a special effort should be made to clear the skin lesions

4 COLLAGEN DISEASE (lupus erythematosus disseminatus, periarteritis nodosa, dermatomyositis, scleroderma).—Here we must indeed be wary, for these conditions can and do almost exactly simulate early rheumatoid arthritis. Fortunately they are not common and the differential diagnosis can be made if one is alert to the possibility. Multiple system

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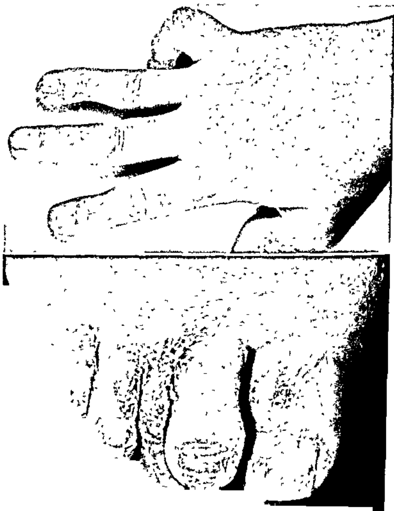


FIG 31 *(top)* —Psoriatic arthritis Note the terminal joint of the fourth finger and extension to the proximal joint—typical of the disease
FIG 32 *(bottom)* —Toenail involvement and extension to the joints

volvement is the clue to collagen disease When a patient ith minimal joint changes who is systemically ill presents ne or more of the following changes, suspect collagen disease: *(a)* multiple system involvement, such as pleural eff... ...

...ppears to be ordinary arthritis, of new symptoms ot related to the joints. (For details see Chapter 7 on colgen disease.)

5 DEGENERATIVE JOINT DISEASE (osteoarthritis) —When reumatoid arthritis and degenerative joint disease occur in elatively pure form, there is little possibility of confusion egarding the diagnosis because of the pronounced differnces between the diseases (see p. 60 for diagnostic points n degenerative joint disease) One should remember, howver, that late in rheumatoid arthritis or in the elderly individual it is quite possible for rheumatoid arthritis and degenerative joint disease to coexist In this situation it is almost always the rheumatoid arthritis that is disabling and deserves the most attention therapeutically "Mixed arthritis" is a poor term but is often used with reference to this condition It is better to diagnose rheumatoid arthritis and degenerative joint disease

6 GOUT.—Early intermittent gout is most often confused with atypical rheumatoid arthritis, i e , palindromic rheumatism (see p 83 for differential diagnosis of gout) Chronic gout is frequently confused with chronic rheumatoid arthritis Although extremely rare, it is possible for a patient to have both gout and rheumatoid arthritis We have followed several such bona fide cases for a number of years

7 SHOULDER-HAND SYNDROME —This distressing condition

may exist either unilaterally or bilaterally. It is characterized by pain and limitation of motion in the shoulder and pain and often swelling and limitation of motion in the hand and fingers. Atrophy of the muscles and demineralization of bone occur rapidly. This condition, because of the pain and swelling of the fingers, is frequently diagnosed as rheumatoid arthritis. It is commonly associated with, or follows, one of the following conditions, although it may develop without known cause. (*a*) myocardial infarction, (*b*) pulmonary infarction, (*c*) cervical cord injury or disease, (*d*) cerebrovascular accident, (*e*) fractures or trauma.

The mechanism of production is not clear, but it is thought to be a reflex nerve disturbance. The course is variable. Patients with mild cases usually recover completely on a program of simple corrective exercises and assistive motion, particularly with regard to restoration of shoulder motion. Severe cases are exceedingly chronic and stubborn problems, but they do not progress as rheumatoid arthritis. The swelling in the hands is usually diffuse and *not limited to the joints alone*. There is often a tendency to the development of fibrous contractions in the palmar area, similar to Dupuytren's contractions. The sedimentation rate is seldom elevated.

No treatment has been uniformly successful. In our experience, the most important aspect of treatment is maintenance of shoulder motion with the maximal amount of rest in abduction, which tends to reduce the edema and swelling. There is no substitute for careful, daily, assistive and stretching exercises supervised by a competent physiotherapist. Baking and massage have not been effective in our hands. Perivertebral sympathetic nerve or stellate block with

procaine may be helpful in severe and acute cases. Aspirin (60–80 gr. daily), a sleeping tablet and, often, codeine sulfate (½ gr. three to four times daily) will be required during early treatment because the pain is severe.

8 "SUBDELTOID BURSITIS" (subacromial bursitis, periarthritis of the shoulder, supraspinatus tendinitis, etc) —See page 74, for a discussion of this condition

9 GENERALIZED FIBROSITIS.—There should be no confusion between this condition and rheumatoid arthritis owing to the absence of joint swelling and the lack of any confirmatory evidence of joint disease from physical, x-ray and laboratory examinations (See p 77 for discussion of generalized fibrositis.)

10 SPECIFIC INFECTIONS —*a)* Gonococcic arthritis. Since the advent of sulfonamides and penicillin, the occurrence of gonorrheal arthritis has been tremendously reduced and is now seen quite rarely Postgonorrheal rheumatoid arthritis, which is seen not infrequently, is most commonly misdiagnosed as gonorrheal arthritis The dramatic response to penicillin, culture of gonococci from the joint fluid, the low glucose content of the joint fluid (average approximately 0 30 mg. per 100 cc) and the x-ray appearance usually permit one to make the diagnosis quite accurately.

b) Brucellosis This does not commonly cause persistent swelling in any one joint and seldom should be mistaken for rheumatoid arthritis The history of exposure, the severe sweats, the rising agglutination titer and positive cultures for brucella should define this condition clearly

c) Pneumococcic arthritis, tuberculous and other septic joint diseases. These are usually monoarticular and easily differentiated

11. REITER'S SYNDROME.—This name has been attached to the symptom complex of urethritis, conjunctivitis, arthritis and sometimes enteritis. Nothing is known with certainty regarding its etiology. It is largely a self-limited disease with spontaneous recovery in three to six months and few or no joint sequelae It has rarely been reported in civilian practice Streptomycin is said to be sometimes effective.

12 STILL'S DISEASE (JUVENILE RHEUMATOID ARTHRITIS).—There is no doubt that the symptom complex spoken of as Still's disease is, in fact, juvenile rheumatoid arthritis. There are two quite striking differences between the prognosis in juvenile rheumatoid arthritis and that in adult rheumatoid arthritis Mortality from acute juvenile rheumatoid arthritis is high, whereas that from the adult type is very low. The joint disease usually is not so destructive in juvenile rheumatoid arthritis and frequently, if the full basic routine (p. 39) is followed in therapy, the outlook is reasonably good. Spontaneous remissions often occur at or following puberty. Many children recover almost completely and go on for years in young adult life without serious sequelae or limitations because of the disease. The prevention of deformity and the maintenance of joint motion and muscle tone are of the utmost importance in this group. (See Chapter 9 for prevention of deformity)

TREATMENT

Most therapeutic efforts in this unhappy disease have been empiric and have centered about whatever concept of etiology the particular physician happened to hold Theories of etiology have come and gone more or less in cycles.

The focus of infection theory provided a remarkably wide range for energetic removal of foci Teeth, sinuses, tonsils, gallbladders, appendixes and prostates were attacked with enthusiasm When the supply of removable foci was exhausted and the patient had not improved, vaccines were developed in the hope of producing immunity to those foci which were not removable. Bacterial allergy was then implicated and desensitization tried after failure to produce immunity to the disease by vaccines Circulatory disturbances were investigated and indicted because of certain changes described in the nail capillaries and the common finding of cold and clammy hands and feet This was followed logically by the use of vasodilators, like histamine Intestinal toxemia has long been considered a cause of the disease, and all physicians are familiar with the attempts to reduce the absorption of so-called "toxins" from the gastrointestinal tract by means of colonic irrigations, intestinal antiseptics, purging and the implantation of Bulgarian bacillus. There are those who believe that neuropsychiatric factors have a major etiologic significance in this disease, and it cannot be denied that treatment directed to this concept has been helpful though not curative

Some years ago the endocrine systems came in for a period of enthusiastic attention Menopausal arthritis appeared in the literature and every type of hormone known at that time was used therapeutically We are now in a period of revival of interest in the endocrine effects, as indicated by the reports of administration of large doses of sex hormones in the treatment of this disease The metabolic concept of etiology is a tremendously wide field but not too well defined, and worthwhile investigations have been ex-

tremely scanty. Some years ago, because the finger-nails of patients with rheumatoid arthritis were found to contain less sulfur than normal, a deficiency in sulfur was suspected, resulting in the administration of sulfur injections to large groups of people. Carbohydrate metabolism was thought to be faulty, hence the rigidly low starch diets. Considerable increased evidence in recent years has led to the belief that rheumatoid arthritis is essentially a disease of a disordered intrinsic metabolism, and much research work is being done in this field. Actually, it is a sad commentary that after all these years of guiding our therapeutic efforts along proposed etiologic lines, we must admit that, so far as a single specific etiologic factor is concerned, we are as much in the dark as we ever were. Rheumatoid arthritis thus remains a disease without identification of a specific etiologic agent and without a single specific cure. Ironically enough, as will be seen shortly, the measures known to influence the disease favorably have resulted from more or less accidental observations, and the mechanism by which they act is unknown.

Because we have no specific cure, most physicians have either become therapeutic nihilists or have contented themselves with trying one therapeutic procedure after another without recognizing the necessity of a really adequate basic program of management. Regardless of our hopes for a specific cure which may come in the not too distant future, rheumatoid arthritis in the meantime will continue to develop progressive crippling deformity. *It therefore seems especially appropriate at this time to emphasize the necessity of establishing every patient with rheumatoid arthritis on a basic routine as the first step in the successful management of the disease.* Any specific therapy that will be devel-

oped will not restore destroyed joints, therefore any help that a future cure may provide your patient will depend on how well you maintain him on a basic program of treatment. Such a basic routine must include specific directions by the physician on at least the following eight problems

Basic routine.—This is to be followed for every patient with rheumatoid arthritis

1 Psychologic adjustment is essential for every patient We do not necessarily refer here to professional psychiatry, but we do urge that when the diagnosis is made the physician tell his patient frankly and honestly what is known about the disease and what is not known about it No false hopes should be given and no quick cures can be promised The physician's failure to place honestly and thoroughly before the patient the problem he is facing with rheumatoid arthritis is responsible for more patients consulting quacks and quick cures than any other single factor Adjustments with family, children, business and economic status are also essential. Instead of having the problem explained and a full basic program outlined, patients are too frequently told either that nothing can be done for the disease, or "We will try these new injections for a while and see if they help"

2. Rest and activity should be carefully balanced on the basis of fatigability and pain Moderate activity is helpful provided the patient is not attempting to walk on bent and painful joints and provided the activity is followed by adequate rest (See Chapter 9 for details)

3. Diet and bowel management must be understood by the patient and prescribed accurately by the physician This is essential not because there are special diets which will cure arthritis but to make certain that the patient is on

a highly nutritious, well balanced diet; and unless one is prescribed for him he will most certainly at one time or another embark on one of the fad diets. Diets in general, so far as caloric intake is concerned, should be adjusted to improve nutrition and weight if malnutrition is present and to achieve a more nearly normal weight if the patient is overweight

4. Correction of anemia is essential. The anemia of rheumatoid arthritis is the hypochromic microcytic type and is often stubborn in its response to the ordinary treatment with iron, liver, folic acid or vitamin B_{12} When these agents fail to restore a moderately satisfactory blood picture, transfusions should be resorted to in an effort to maintain the desired blood level.

5. Cautious removal of foci of infection should be undertaken, not with the promise of curing the disease but to improve the patient's general health. Foci of infection in a patient with active acute rheumatoid arthritis should be removed only after careful preparation and preferably after one or two blood transfusions. It is also well to give prophylactic doses of a sulfonamide or penicillin before and after removal of the foci because occasionally removal of foci without these precautions produces a violent exacerbation of the disease.

6 Corrective and group muscle postural exercises should be taught carefully and carried out meticulously because they offer both the ambulatory and the bedfast patient an opportunity to maintain muscle tone and joint motion (See the list of exercises, p 145).

7. Relief of pain is usually quite easily accomplished by the administration of aspirin, salicylates, gentle warmth.

splinting and support of painful joints and other simple cedures. If opiates are required, the diagnosis should questioned.

8 Prevention and correction of deformity are essent In every patient with rheumatoid arthritis, the probabil of residual deformity in all involved joints should be antic pated and prevented This problem is so important th Chapter 9 has been devoted to it.

Elective procedures.—Once the patient on the basic pro gram of management is established, one is justified in looking about for additional measures that may influence the arthritis favorably The list of so-called cures for rheumatism and arthritis runs into the thousands, and their very numbers are good evidence of the lack of specificity. Among the more commonly used agents which in our experience have been of doubtful value or worthless are vaccines, neostigmine, procaine intravenously, fever therapy, ACS (Bogomolets' serum) and gadget physiotherapy Short wave diathermy, whirlpool baths, paraffin packs, ionization and many other so-called physiotherapy methods are expensive and for the most part worthless in the treatment of rheumatoid arthritis It is our deep conviction that the most valuable physiotherapy for the patient with rheumatoid arthritis is the raining of his own muscles through carefully directed exerses (see Chapter 9) These exercises may be done in ater or on the bed, but trained personnel is required to ch and supervise the patient's own activities He must be taught to use his own muscles Warmth is sometimes helpful, but the intensive baking and massage that are so popular, though perhaps comforting at the moment, will keep the disease active and do more ultimate harm than good

The basic program already described is essential for every patient with rheumatoid arthritis. The following elective procedures unfortunately are not applicable all at once or to all patients, but may be selected when suitable and should be given consideration from the standpoint of newer concepts in etiology and the possibility that they will ultimately prove the ideal treatment.

1. TRANSFUSIONS.—Patients with acute or subacute rheumatoid arthritis often respond well and sometimes dramatically to repeated transfusions of ordinary blood. Transfusions are, of course, also valuable in correcting anemias. We have used in excess of 100 pregnant blood transfusions in patients with active rheumatoid arthritis. The results were variable. A few patients who had failed to improve materially after usual blood transfusions made rather spectacular improvement after transfusions of pregnant blood. There were, however, failures with the pregnancy transfusions as well Obviously, transfusion of pregnant blood is not a practical method of therapy and has been done by us only as a research problem in studying the mechanism of the remission when it occurs. Our practice is to prescribe ordinary blood transfusions, one weekly, preferably approximately 300 cc, for three to five or six weeks in acute or subacute cases.

2. GOLD THERAPY.—The use of gold therapy for rheumatoid arthritis dates back approximately 20 years and, despite the hazards, has increased during this period. There is no question that 40–60 per cent of patients who can take adequate gold therapy will show improvement. A smaller percentage may show a complete remission of the disease. Much is written about the therapy of rheumatoid arthritis with gold, but it must be remembered that *rheumatoid arthritis*

of peripheral joints is the only form of rheumatism in which it should be used Rheumatoid spondylitis is not usually benefited. Gold may be toxic, and the most careful supervision is required for successful administration. (For details of gold therapy, see pp 45 f)

3 CLIMATE.—A small percentage of patients with rheumatoid arthritis experience dramatic improvement with a change to a warm dry climate The majority have no striking change, but are usually more comfortable and with equally good medical care are apt to improve more rapidly than they would in less favorable climatic surroundings The chance, however, that a change of climate will help does not warrant disruption of the economic security of a family Climate consists in complex meteorologic factors such as stability of barometric pressure, humidity, altitude, amount of [illegible], concentration of [illegible] temperat[illegible] have found it impossib[illegible] to predict which patients with rheumatoid arthritis will be clearly benefited by climate. If a change of climate is to be considered, three to six months should be planned as a minimal trial period Patients need medical supervision at all times. The advice frequently given patients, "Just go out and bake in the sun," is extremely dangerous Each year patients are seen in delirium from following this advice A systemic reaction on exposure to the sun in dry desert regions can occur without skin burn to guide the length of exposure.

A change of climate may also entail other considerations: absence from home and business, a scheduled routine of living, time available to make a job of getting well, etc Many of these are important in recovery.

4. X-RAY THERAPY.—This is undoubtedly of value i treatment of rheumatoid spondylitis but does not a to be beneficial in the peripheral form of arthritis.

5. PREGNANCY.—Although some would doubt the a ability of including pregnancy among the therapeutic a for rheumatoid arthritis, it nevertheless has a profoun fluence on the disease. Of 96 women with active rheum arthritis observed during pregnancy by us and other per cent showed a remission of the disease during the nancy. We have in a number of instances advised pregn in women with active progressive arthritis which had been altered by other available therapeutic aids. The di is apt to recur following childbirth, but if special ca given the patient and includes rest, small doses of th extract and a few small blood transfusions, remission often maintained for several years. Pregnancy is menti not so much because of its practical applicability but cause, like jaundice, of its strong antirheumatic action.

6. JAUNDICE.—This is not available by clinical pres tion, but it is well known that an adequate attack of jaun may produce a striking reversal of rheumatoid arthritis.

7. CORTISONE AND ACTH.—Cortisone is a steroid horn produced by the adrenal cortex. It is at present manufact in very small quantities and by a long series of chem processes from the steroid nucleus in bile. ACTH (adr corticotrophic hormone) is produced by the anterior p tary gland and regulates the activity of the adrenal co This substance is secured at present in very small quant from the pituitaries of hogs. These two substances are newest and most powerful antirheumatic agents so far served. Since the initial dramatic report of Hench, Ken

Slocumb and Polley, of the Mayo Clinic, indicating that these substances were capable of reversing the process of rheumatoid arthritis, it has been clearly established by us and others that these substances do produce a striking remission and a reversal of the active disease in patients with active rheumatoid arthritis. (See Chapter 8)

These very observations that rheumatoid arthritis, our most crippling disease, can be completely reversed and the patient returned to normal under the circumstances of pregnancy, jaundice, gold, cortisone, ACTH therapy, and *sometimes transfusions, offer us the brightest possible hope* that we shall discover the mechanism of the remission and that further research will uncover a certain and specific treatment and perhaps preventive for rheumatoid arthritis Contrast this with the present irreversibility of cancer, infantile paralysis and chronic heart disease Advances are being made in research in all the rheumatic diseases The greatest danger lies in our tendency to put off doing the basic things required for the patient while waiting for the quick cure. The entire field of rheumatic diseases can be understood and reasonably accurate diagnosis can be made by any physician who will make the effort and spend a reasonable amount of time on the problem Treatment of rheumatoid arthritis has not yet been reduced to specific therapy, but an honest attempt to treat adequately one's patients with this disease will yield a far greater degree of success than is generally supposed.

GOLD THERAPY

Indications.—Gold therapy is much more successful in early rheumatoid arthritis without much joint destruction

than in the late stages with destroyed joints. After the basic routine (p. 39) is established, if progress is not made in the first month or two, one may consider the advisability of gold therapy. When the patient's organic status is suitable, except for the arthritis, the risk of gold therapy may be less than that of progressive, uncontrolled joint damage, and gold therapy may be indicated. Results in more than 2,000 cases, collected from the literature and our own experience, indicate that one may expect about 60 per cent of the patients who can take gold without having toxic reactions to show definite improvement. A smaller percentage may show complete remission.

Preparations.—There are six or more gold compounds on the market, including gold sodium thiosulfate (Sanocrysin), gold sodium thiomalate (Myochrisyne), gold thioglucose (Solganol-B oleosum), gold sodium succinimide and gold thioglycoanilid (Lauron) Of these products, gold sodium thiomalate and gold thioglucose have been most widely used in this country The gold content of the various salts varies from approximately 37 to 54 per cent. Their solubility also varies, gold sodium thiomalate being insoluble and gold thioglucose soluble in water; our experience has been largely with gold thioglucose and gold sodium thiomalate.

Dosage—The routine is 10 mg. of the soluble salt twice the first week and 25 mg. weekly for three weeks thereafter. If the patient is small, the dosage will remain at 25 mg. once weekly. If the patient weighs over 120 lb., it is our custom after three weeks at 25 mg. to increase the dose to 50 mg. weekly. When deciding to use gold therapy one must weigh the possible hazards of toxicity against the serious outlook of the disease.

We do not use the large doses of 100 mg. weekly because of the pronounced increase in toxic symptoms experienced The dosage of 50 mg. weekly should be continued for three to five months at least, provided no toxic symptoms appear, before deciding that gold will not be effective If remission or striking improvement occurs and the sedimentation rate drops during this period, the interval of dosage is increased so that the patient will receive 50 mg. each two weeks With satisfactory improvement in the clinical and blood pictures on this routine, the interval may be increased to once in three weeks Because of the hazards of exacerbations and recurrences of the disease when gold therapy is interrupted, we no longer give gold in courses and stop when a specified amount has been given Many patients who have had excellent remissions on gold may have severe exacerbations a few months after therapy is stopped, and these patients often do not respond so readily to retreatment with gold. Our experience suggests that, once having attained an excellent result and a remission of the disease, maintenance doses every two to three weeks should be continued for at least a year or two and perhaps longer

Toxic reactions.—The mortality rate calculated in over 2,000 cases was 0 39 per cent. Toxic reactions of some kind occurred in 32 per cent of this group. The higher the weekly dosage, the more apt the patient is to show toxic effects. The toxic reaction most frequently encountered is dermatitis. This may be of any degree, from a mild rash to a severe exfoliative type of dermatitis. Other toxic reactions include ulcerative stomatitis, hepatitis, nephritis, ulcerative colitis, thrombocytopenic purpura, agranulocytosis, aplastic anemia, cerebral involvement and bronchitis. Such

toxic reactions usually are encountered after several hundred milligrams of the drug have been given, but occasionally they are seen following the initial small dose. Toxic reactions may also manifest themselves weeks after the treatment is completed There is no definite, sure way of preventing such reactions. It is possible that liver injections and administration of the vitamin B complex, vitamin C and antihistamines and other general measures are helpful, but they are in no way specific for prevention of toxic reactions to gold.

Treatment of toxic reactions.—Because of the overwhelming evidence that British anti-lewisite (BAL) was effective in treatment of arsenical and mercurial poisoning, it has been tried in the treatment of gold toxicity by a number of investigators. Many of the toxic reactions to gold are extremely mild and self-limiting on discontinuance of the gold. BAL is an effective antidote to the gold toxicity, if given early. If the gold reaction is severe, either a severe dermatitis or one of the other serious evidences of toxicity, BAL should be given early and in adequate doses. One hundred milligrams should be given intramuscularly as the initial dose and, if there is no unfavorable reaction, followed four hours later with 200 mg. and, again if no serious reaction occurs, repeated to a total of 800–1,000 mg. during the first 24 hours. The duration of treatment depends on how rapidly the evidence of toxicity clears. BAL itself may produce some toxic reactions, occasionally causing urticaria, abscesses at the site of injection and nausea and vomiting. We have found ACTH highly effective in blocking or relieving the skin manifestations of gold toxicity; so, if available, it is the treatment of choice.

Contraindications to gold therapy.—Gold therapy should

be avoided in the presence of severe diabetes, nephritis, ulcerative colitis, hepatic insufficiency, blood dyscrasias, a history of exfoliative dermatitis, rheumatic fever and disseminated lupus There are undoubtedly other contraindications

Precautions in use of gold.—1 Before giving each injection, look for rash, purpuric spots, stomatitis, albuminuria, microscopic hematuria and any unusual symptom which the patient may report

2 Check every two or three weeks for leukopenia, low platelet count and changes in the hemoglobin content and differential count

3 Do not give gold unless you are familiar with the bad results It is our conviction that this is one injection which the physician should give and not delegate to a nurse.

CHAPTER 3

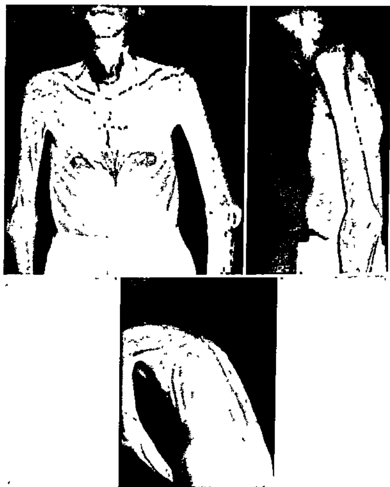
Rheumatoid Spondylitis

RHEUMATOID SPONDYLITIS is also known as Marie-Strümpell disease, spondylitis ossificans, spondylitis ligamentosa, ankylosing spondylitis and spondylitis rhizomelica. This form of rheumatism is important because it is the most frequently misdiagnosed condition among the rheumatic diseases. The failure to diagnose it properly is due almost entirely to failure to consider it as a possibility, because the diagnosis is quite easy once it is thought of. There is wide difference of opinion as to whether rheumatoid spondylitis is actually rheumatoid arthritis with a predilection for the spine or whether it has some additional variant and should be considered a separate disease. There are three arguments for considering it a separate disease. (1) The sex incidence is approximately 9 males to 1 female, whereas peripheral rheumatoid arthritis roughly involves 3 females to 1 male. (2) Gold therapy is not effective, whereas in peripheral rheumatoid arthritis it may be effective. (3) X-ray therapy is effective, whereas in peripheral rheumatoid arthritis it is not. Two arguments favor its classification as rheumatoid arthritis. (1) In approximately 25 per cent of the patients peripheral joint changes develop that are indistinguishable from those of rheumatoid

arthritis. (2) The pathologic changes in the lateral articulations of the spine in rheumatoid spondylitis are similar if not identical to those in the peripheral joints in rheumatoid arthritis.

In its late stages rheumatoid spondylitis is so characteristic that the diagnosis is obvious. The young man with a stiff back who tends to protrude the head and develop wagon-wheel deformity, walking with a waddling gait, often with hip and shoulder limitation, sometimes with fixed distortions of the pelvis or spine, presents the late picture of this disease (Figs. 33–35). The onset may be acute (15 per cent) or insidious (85 per cent) The early symptoms are nearly always misdiagnosed Frequently fibrositis, lumbago, lumbosacral strain, sciatica or muscular rheumatism is diagnosed in the early insidious type of onset.

In a high percentage of cases the first complaints are referred to the lower part of the back and consist of periods of aching or stiffness, transient sciatica and difficulty in bending forward to pick up objects from the floor In some cases these symptoms are intermittent, with complete freedom between attacks for a number of months or years In others the symptoms continue to increase slowly and the patient begins to notice that coughing, sneezing, turning himself in bed or being jarred during a ride over a rough road produces pain, often low in the back and frequently in the rib cage General constitutional symptoms develop, of which weight loss is the most constant and dramatic. Many patients lose from 20 to 60 lb. in a few months or a year. There are fatigability, loss of appetite, sometimes low grade fever. During this stage ordinary anteroposterior and lateral x-rays of the lumbar region of the spine usually reveal no evidence of



FIGS 33–35 —Rheumatoid spondylitis. Late effects, with peripheral joint involvement, malnutrition, flexion deformity of the spine, fixed rib cage and striking muscle atrophy.

disease, but x-rays of the sacroiliac joints will almost always reveal changes that are characteristic of the disease (Figs 42–45, p. 57). As the disease progresses upward from the lumbar region to the dorsal and cervical areas, flattening of the chest and fixation of the costal cage occur Vital capacity is markedly diminished. Because of the collapse and fixation of the costal cage, the head tends to pull forward and the neck develops a characteristic deformity and loses motion The deformity may start in flexion and often goes into hyperextension, compensating for the flexion in the dorsal region of the spine The shoulders and hips are frequently involved, with gradual decrease in motion, and flexion deformity is likely to develop in the hips Early diagnosis is essential, for it is only then that treatment can prevent these deformities

DIAGNOSIS

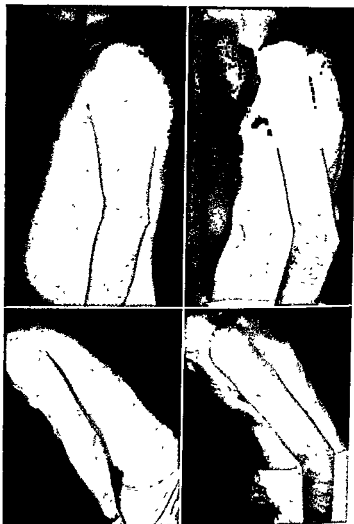
The following observations should lead to correct diagnosis

1. Rheumatoid spondylitis should be suspected in any young man who has recurrent low back pain, sciatica or lumbago.

2 Excessive loss of weight with vague symptoms of muscle or joint pain should suggest rheumatoid spondylitis

3 Pain low in the back or in the chest on coughing or sneezing and difficulty in turning over in bed are typical symptoms of rheumatoid spondylitis

4. Limitation of motion of the lumbar spine and flattening of the lumbar curve usually occur early (Figs. 36–39) There may also be limited chest expansion (Fig 40) Tenderness and spasm of lumbar muscles are usually present



FIGS. 36 AND 37 *(left top and bottom)*—Rheumatoid spondylitis, early stage. Appearance is normal until flexion of the spine is attempted, when *straight back* is easily noted

FIGS 38 AND 39 *(right top and bottom)*—Moderately advanced stage Some deformity is apparent in upright position; the lumbar curve is flattened on flexion.



Fig 40 (*top*) –Rheumatoid spondylitis The chest looks normal, but maximal change from forced expiration to forced inspiration was only ¼ in, indicating fixation of the rib cage

Fig. 41 (*bottom*) –Chest compression sign It is positive when dorsal vertebrae are involved It may be elicited early, before there is much limitation of the rib cage

CHAPTER 4

Degenerative Joint Disease

DEGENERATIVE JOINT DISEASE, or osteoarthritis or hypertrophic arthritis, is actually not so much a disease as it is a wearing out of tissue. The manifestations may be observed in either of two forms.

1 The localized form may occur at any age and is secondary to major trauma, faulty structure or previous joint infection. The symptoms are localized and usually the diagnosis is easily made on the basis of the history and clinical examination and roentgenographic studies of the affected joint.

2 Generalized degenerative joint disease often involves the spine, knees and hips or other weight-bearing joints Heberden's nodes (enlargement of the end finger joints) occur frequently; they have been shown to have a familial tendency. This type of arthritis occurs usually in older people and has been called "old folk's rheumatism" The exact cause of the degenerative process is not known; however, some of the chief contributing factors are overweight, faulty posture and weak muscle tone, all of which produce constant "minor trauma" and inevitably lead to extra stress and strain on the joints

DIAGNOSIS

The diagnosis of generalized degenerative joint disease is not difficult. The following points are helpful in arriving at the diagnosis.

1. The spine and weight-bearing joints are usually involved.

2 The end joints of the fingers are often enlarged (Heberden's nodes) (Figs. 46–48).

3. *Onset is insidious*

4 Symptoms first appear in persons past middle age.

5 The patient is often overweight and has a low basal metabolic rate

6. *There is seldom much soft tissue swelling* such as is present with rheumatoid arthritis The swelling in degenerative joint disease usually feels hard and firm in contrast to the soft fluid swelling of rheumatoid arthritis.

7 Usually x-ray studies show increased mineral deposits with sharpened articular margins Figures 49–56 show normal joints and joints affected by degenerative disease. There may be eburnation of subchondral bone and joint cartilage destruction. When narrowing occurs in the disk spaces between the vertebrae, lipping of vertebral margins is common.

8. The disease seldom affects the wrists, feet or metacarpophalangeal and proximal interphalangeal joints, which are so often involved in rheumatoid arthritis.

9. The sedimentation rate is normal or slightly *elevated*

10. The patient usually is not systemically ill.

11. Symptoms chiefly complained of are pain on motion of or weight-bearing by the involved joints, stiffness of the *back or knees on getting up from a chair* and pain or swelling in the end finger joints





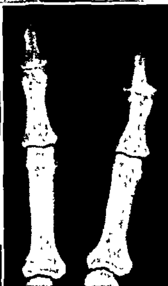


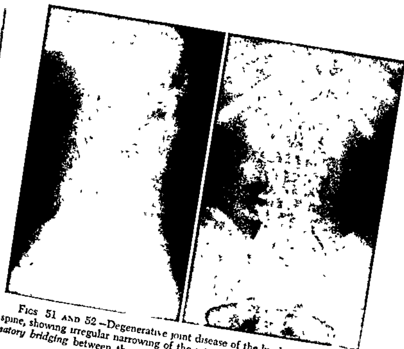
Fig 46 *(top)* —Degenerative joint disease (osteoarthritis) Note enlarged end joints (Heberden's nodes)

Fig 47 *(bottom left)* —Heberden's node

Fig 48 *(bottom right)* —X-ray of same joint




FIG. 49 *(left)*—Normal lumbar region of the spine.
FIG 50 *(right)*—Normal lumbosacral joint.



Figs 51 and 52 — Degenerative joint disease of the lumbar region of the spine, showing irregular narrowing of the intervertebral spaces and compensatory bridging between the vertebrae



FIG 53 *(top left)* —Normal knee, anteroposterior projection

FIG 54 *(top right)* —Degenerative joint disease (osteoarthritis of the knee, anteroposterior projection Note sharpening of the spines of the tibia, in contrast with the normal

FIG 55 *(bottom left)* —Normal knee, lateral projection

FIG 56 *(bottom right)* —Osteoarthritic knee, lateral projection Note good mineralization and patellar spurs

Three types of degenerative joint disease deserve special mention

1. *Involvement of the hip (malum coxae)* may be extremely disabling Pain in the hip may be the first symptom, often with early loss of extremes of motion. There are cartilage destruction, with narrowing of the joint space, and deformity of the head of the femur, and the disease is progressive, with increasing limitation of hip motion and severe pain on weight-bearing One or both hips may be involved Symptoms may be limited to the hips or the hip involvement may be only a part of widespread joint disease The x-ray picture is characteristic (Figs 57–59) Treatment is the same as that outlined for generalized degenerative joint disease (pp 71 ff)

2 *Osteoarthritis of the spine* is the second type of degenerative joint disease worthy of special mention This diagnosis is frequently made to account for low back pain Many such diagnoses are made only because of low back pain and x-ray evidence of lipping or spurring of the vertebral margins Osteoarthritis of the spine is not as common a cause of pain as the frequency of this diagnosis would suggest X-ray evidence of spurs or hooks (osteophytes) on the anterior margins of the vertebral bodies is not uncommon in patients who have no pain or symptoms (Figs 51 and 52) These spurs can and do occur as well in patients who complain of back pain The osteophytes are merely calcified annular ligaments and represent in a sense the tombstones which mark the site of loss of disk substance It is true that loss of outline of disk material may result from degenerative joint disease of the spine (osteoarthritis) or from unrelated etiologic factors such as trauma and infection The sole pa-





FIG 57 *(top)*—Unilateral malum coxae, with loss of hip joint cartilage and deformity of the femoral head

FIG 58 *(bottom left)*—Early malum coxae, showing loss of hip joint cartilage but little demineralization, cyst formation or deformity of femoral head

FIG 59 *(bottom right)*—Far-advanced malum coxae, showing almost complete obliteration of the joint space with areas of demineralization, and of condensation with deformity of the femoral head



FIG 60—Degenerative joint disease of cervical vertebrae, with narrowing of the intervertebral spaces and hypertrophic marginal changes

tain percentage of cases of low back pain. It is well to remember that backache and neck pain occur far more frequently from soft tissue (muscle and tendon) disturbances than from osteoarthritis. The single observation that degenerative joint disease (osteoarthritis) is present should seldom be accepted as an adequate explanation for symptoms. Pain in the neck associated with occipital headache is often ascribed to osteoarthritis of the cervical vertebrae. Such a condition can, of course, cause pain, but the symptoms are far more frequently due to fibrositis of the neck muscles (see p. 76).

3. *Senile osteoporosis* (Figs. 61–63) is often not diagnosed. There is marked decalcification which, in the spine, frequently results in the collapse of one or more vertebrae. The patient may note a loss of several inches in height. Nerve root involvement with radicular pain is common. The cord may, of course, be injured if vertebral disintegration is sufficient. The treatment includes corrective exercises, back brace, if needed, and administration of male and female hormones. It is our practice to give women (usually over 60) methyl testosterone in 5–10 mg doses daily, in the form of sublingual or buccal tablets. It is given for three weeks, followed by one week of diethylstilbestrol in doses of 0.25–0.5 mg daily. If side-effects of the methyl testosterone (sexual awareness, slight edema, weight gain and husky voice) become unpleasant, the drug should be discontinued before the three week course is completed and the one week course of diethylstilbestrol begun. The methyl testosterone should be started again at the end of the week's treatment with diethylstilbestrol and continued for three weeks, or until side-effects become unpleasant, thus alternating the two



Fig 61.—Normal lumbar region of the spine



Fig. 62.—Early senile osteoporosis



Fig 63.—Well established senile *osteoporsis*

medications for six months or more Occasionally even quite elderly women have slight vaginal bleeding while taking the female hormone. The patient should be warned of this possibility and instructed to stop the female hormone and return to the methyl testosterone should bleeding occur We prefer to administer the hormones separately rather than together because we wish to give the minimal amount of female hormone to these elderly women, who are in the cancer age

TREATMENT OF GENERALIZED DEGENERATIVE JOINT DISEASE

A positive diagnosis, intelligent advice and assurance that the disease is not crippling will do much to dispel the anxiety of the patient (especially a woman) with Heberden's nodes and painful knees Reassurance is one of the most effective aspects of therapy of this disease Treatment in general should be directed toward relieving the excessive stress and strain resulting from constant minor trauma to the joints and alleviating the "wear and tear" factors The following suggestions will be helpful

1 After the patient has been reassured, insist on the establishment of a routine, depending on the degree of the symptoms Dismissal of the patient with the mere assurance that the disease is not serious or crippling will result in his going elsewhere for relief of his discomfort

2 Weight reduction if the patient is overweight

3 Correction of posture

4 Routine daily corrective exercises for strengthening the supporting muscles (See the list for rheumatoid arthritis, p 145, and select suitable ones)

5. Shoe correction, for better weight-bearing angle.

6. Achieve a balance between rest and exercise which the patient can tolerate, and gradually increase the activity as weight is reduced, posture is improved and muscle strength is increased.

7. Prescribe aspirin, which is the most helpful drug generally for the relief of pain and stiffness. It should be used in adequate dosage (30–60 gr. daily).

8 Short periods of warming and light massage of the muscles are soothing but not essential.

9. Application of Ace bandages for support of a painful knee until muscle tone can be improved is often helpful in walking (see pp. 131 and 135).

10. Insist on the use of crutches whenever there is hip involvement or severe knee pain. Correct crutch walking should be taught (Figs 100 and 101, p. 132). This maintains correct posture and balance and prevents excessive stress on the painful weight-bearing joints.

11 Intravenous or local injection of procaine, curare, x-ray therapy, sex hormone therapy and many other procedures are occasionally helpful but usually not essential

12 General maintenance of warmth and protection, use of electric blankets, etc., add much to the patient's comfort

13 Head traction with a Sayre head sling for 20–30 minutes, combined with heat and gentle rotation of the head, is helpful when the cervical spine is involved Weight should not be over 20 lb. to start and is gradually increased as tolerated

With the exception of degenerative joint disease of the hips (malum coxae), this condition is essentially benign and can be described as not the "crippling kind" of arthritis. Pa-

tients will in general respond satisfactorily to an adequate treatment program such as that described here, which is neither complicated nor expensive

CHAPTER 5

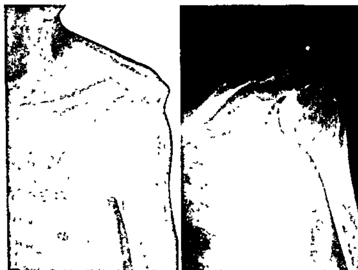
Fibrositis

FIBROSITIS, OR "muscular rheumatism," is said to be the most common form of chronic rheumatism. There are two general types.

1 *Localized fibrositis* may result from injury, infection or exposure or exist in association with arthritis around a joint This type is usually not difficult to diagnose and is ordinarily quite easily and successfully treated. Treatment should be directed to protection of the injured part, removal of infection, avoidance of exposure, use of selected exercises, compresses, intermittent heat and, in the late stages, massage In many instances, the infiltration of 10 cc. of 1 per cent procaine into the painful area permits increased motion, relieves pain and hastens recovery. There are several special types of localized fibrositis that deserve mention

a) "SUBDELTOID BURSITIS" (periarthritis of the shoulder, subacromial bursitis, supraspinatus tendinitis, painful shoulder, etc) —This condition is the cause of approximately 90 per cent of painful shoulders. It is frequently encountered, is the cause of excruciating pain and may result in a "frozen shoulder" (Figs. 64 and 65). It is generally believed that the process starts in the tendon of the supraspinatus muscle

and extends to the bursa. This painful condition is almost always due to minor or major trauma, exposure, foci of infection or any combination of these factors It is usually unilateral The severe pain in the shoulder produces acute discomfort, particularly on abduction and rotation Occasionally x-rays disclose a calcified area in the region of the bursa If untreated, this condition may result in a so-called "frozen shoulder" with motion limited to a few degrees There may be secondary discomfort in the hand, but usually not with the swelling, marked atrophy and hand pain that is so common in the shoulder-hand syndrome (p. 33)



FIGS 64 AND 65 —Neglected subdeltoid bursitis with completely "frozen shoulder Note muscle atrophy and rather normal x-ray appearance of the joint

During the acute attack, rest, x-ray therapy to the shoulder (*usually 50 r at 30 cm. distance daily for four days, then* once weekly for two additional treatments) and the maintenance of shoulder motion by a gentle and wise physical therapist will often result in a painless shoulder with free motion. Additional x-ray therapy of the acute attack is seldom helpful if there has been no improvement with the first course. Aspirin (60–80 gr. daily), codeine sulfate (½ gr. three or four times daily) and a night sedative may be required if pain is severe. If the condition is seen late when there is already pronounced shoulder limitation, short wave diathermy followed by patient and skilful assistive and stretching exercises to restore the shoulder motion will usually over a period of weeks or months restore a painless shoulder and secure normal motion. Occasionally, late in the disease, very gentle manipulation under anesthesia is necessary to break adhesions that limit the shoulder motion

b) HERNIATED FATPAD.—This condition occurs most frequently in the back, in the lumbar and lumbosacral areas. Distended lobules of fat, when confined in the normal fibrous integument, may produce pain. A distended lobule may herniate through the fibrous covering and often is easily felt on palpation. Simple heat and massage combined with procaine injection is usually successful. Unless the pain is intractable, surgical removal should not be undertaken.

c) FIBROSITIS OF THE NECK.—Pain in the neck, often associated with headache, is a *frequently* encountered type of discomfort caused by fibrositis in the neck structures. The lymph nodes are usually enlarged and tender, the muscles tight and spastic, and the headache and neck discomfort can be severe enough to be disabling. The condition is usually

associated with foci of infection in the sinuses, nasopharynx or mouth. The patients are extremely sensitive to cold, drafts, exposure and fatigue. On the basis of x-ray findings, such patients often go on for years, receiving treatment for osteoarthritis of the cervical spine, without discovering the true cause of the disability. Figure 66 shows striking osteoarthritis of the cervical spine in a woman, aged 51, who had had severe neck pain and occipital headaches for three years. She had been treated for osteoarthritis with head traction, procaine injections and all the usual remedies, none of which had relieved her. Complete relief from all symptoms followed the clearing up of chronically infected adenoid tissue in the nasopharynx by means of radium application. Osteoarthritis was present, but was not responsible for the symptoms. Figure 67 shows early osteoarthritis that caused severe symptoms. If the fibrositis is of long standing, a long course of heat therapy and vigorous massage may be required. These measures should be undertaken only after a careful survey and removal of infected foci in the head.

2. *Generalized fibrositis*, on the other hand, is an elusive ill-defined term. There are no certain diagnostic criteria. The diagnosis of generalized fibrositis is often used as a "catch basket" for patients who complain of generalized joint or muscle pain, usually without any substantiation from laboratory, x-ray and physical examinations. It is perfectly true that tenderness on pressure will be found in many areas, although no specific pathologic process is identifiable with regularity. It is quite probable that most cases of generalized fibrositis belong in the classification better described as "anxiety state" or "psychoneurosis with skeletal symptoms." Recently the condition in some patients of this group, who

show quite typical psychoneurotic symptoms of the conversion hysterical type with fixed patterns, has been termed "psychogenic rheumatism" These patients should be recognized and given psychiatric treatment. Instead of using the



Fig 66 *(left)* —Pronounced osteoarthritis of the cervical portion of the spine, but not the cause of the patient's symptoms

Fig 67 *(right)* —Early osteoarthritis of the cervical portion of the spine Changes appear to be less extensive than those in Figure 66, yet the patient's symptoms were due to the arthritis

term "psychoneurosis with skeletal manifestations" or "anxiety state" for the balance of this group, we have adopted the expression "tension pain" and find that the patient understands this term quite well.

These patients sit in the office or lie on the examining

table with every muscle held tense. If the patient's anxiety, often unconscious, results in constant muscle tension, the pain may be very real, for continuous tension in the same muscle group will produce pain in a normal person. The typical patient in this category generally complains of pain and aching in the muscles and joints, usually widely distributed, worse in the morning on awakening and improved with exercise. Physical examination usually gives negative results except for tenderness on pressure over many different points and soreness of the muscles. X-ray and laboratory examinations do not add any positive findings Atrophy, swelling, limitation of motion and evidence of constitutional illness are seldom present Such a patient is usually anxious, tense and frightened at the thought of having arthritis It is not uncommon, when one sees them in consultation, to find these patients taking five or six kinds of physiotherapy, numerous injections and oral medications

TREATMENT OF GENERALIZED FIBROSITIS

The patient cannot be dismissed with the assurance that nothing can be found His emotional and tension problems are often obscure, and a little time spent in listening to the patient's story will often pay great dividends in therapeutic results The patient must be given a careful explanation that will satisfactorily account for his pain and the assurance that he can do something about relieving it Regardless of etiology, we are convinced that, given an adequate explanation of their problem and guidance in the use of relaxing and stretching exercises (see list, p 151), together with active exercise, these patients will show gratifying improve-

ment. Many patients will gladly accept such an interpretation of their problem and with sympathetic encouragement will often pour out their anxieties and fears. This mental catharsis in itself has often been strikingly beneficial. Certainly, if an understanding of anxiety and tension pain together with the most superficial psychiatric aid and simple self-help measures will succeed in relieving symptoms, this is a method of treatment to be preferred over the usual gamut of vitamins, vaccines, ionizations and gadget physiotherapy that is usually so unsuccessfully employed in these problem cases. These patients, though lacking in positive physical signs and evidence of organic disease, are usually just as ill and miserable as those who have definite changes They deserve advice on a definite routine of living, wise periodic guidance and an understanding of their problems. Such a course of management will often transform what are known as "office pests" or "crocks" into grateful and happy patients.

CHAPTER 6

Gouty Arthritis

Gouty arthritis is an intrinsic disorder of uric acid metabolism. Hyperuricemia appears to be hereditary, but all indi-

tive in 50–75 per cent of cases The typical life story of a patient with gouty arthritis may show hyperuricemia for years before an attack of arthritis First attacks usually occur in a man at about the age of 45, though they may occur in the twenties and through the sixties. First attacks are usually acute, with joint swelling and pain, often in a single joint, lasting a few days to a few weeks, with complete recovery The attack is usually dramatic and severe and often occurs at night The period of remission or freedom after the first attack may vary from a few weeks to several years Gradually, however, attacks may become more frequent and of longer duration If this cycle continues, chronic gouty arthritis becomes established and there will be no periods of complete remission (Fig 68). This may require 10 years or longer Multiple joints become involved, and the disease may become as crippling as rheumatoid arthritis

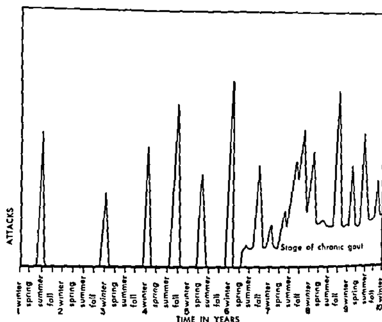


FIG 68 —Typical life history of gouty arthritis.

The exact mechanism of uric acid disturbance associated with the acute attack is not known. Recent investigations suggest that it may be related to a faulty androgen produced by the adrenal cortex.

DIAGNOSIS

The diagnosis of gouty arthritis is not based on the blood uric acid level owing to difficulties in the accuracy of methods and to the fact that many individuals may have a high blood uric acid level without having gouty arthritis. It is quite probable that if all patients with gout were examined frequently by accurate methods, most of them sooner or

later would be found to have elevated blood uric acid levels. Patients may be seen, however, in an acute attack of gout with normal blood uric acid levels. The diagnosis usually can be made quite easily by using the following diagnostic hints.

1. History of acute attacks of arthritis, ordinarily limited to one area, subsiding promptly with complete recovery, usually in a man. The ratio is about 97 men to 3 women. The big toe, commonly thought of as the gouty joint, is involved initially in only 60 per cent of cases. Gout does, however, tend to involve peripheral joints—seldom the hips, shoulders or spine

2. Striking remission by the use of colchicine (see p. 87), given until diarrhea occurs, during or at the onset of an acute attack

3 A tendency for the acute attack to occur after surgery and after trauma (as after a hunting trip).

4 Demonstration of uric acid in the joints (Figs 69 and 71) and in the tophi on biopsy

5. Occurrence of renal colic or the demonstration of renal calculi as corroborative evidence

6 X-rays not diagnostic until the late stages (Figs 70 and 72)

Differential diagnosis —Gout may be confused early in its course with acute rheumatic fever, atypical rheumatoid arthritis, i e., palindromic rheumatism, traumatic arthritis, osteoarthritis and certain rather rare conditions

1. The initial attack of rheumatic fever usually occurs before age 40, whereas the initial attack of gout occurs after age 40. In rheumatic fever there is a migratory arthritis, which is rare in gout, and there is no scaling or peeling of the skin over the joint, as is seen in gout. The colchicine

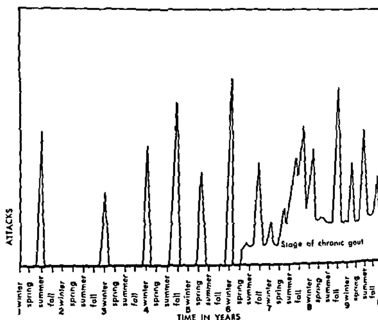


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FIG 71 *(top)*—Chronic tophaceous gout of the knee
FIG 72 *(bottom)*—Same case x-ray of the left knee



FIG 69 *(top)* —Far-advanced tophaceous gout.
FIG 70 *(right)* —X-ray of the same foot





FIG. 71 *(top)*.—Chronic tophaceous gout of the knee.
FIG. 72 *(bottom)*.—Same case, x-ray of the left knee.

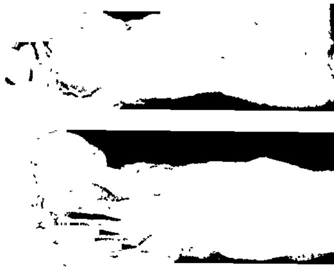


Fig 69 *(top)*.—Far-advanced tophaceous gout
Fig 70 *(right)*.—X-ray of the same foot

2. Protection of joints with cradle, footboard or cotton batting

3 Colchicine (1/100 gr.) every two to three hours, as tolerated, until diarrhea occurs. The diarrhea, if not self-limiting, may be controlled quickly with paregoric. The diarrhea, or therapeutic point, may develop within 24 hours but may be delayed as long as three days If nausea develops before the diarrhea, reduce the frequency of the colchicine administration. Definite diarrhea (three or four watery stools) is the best guide to adequate dosage An acute attack of arthritis not relieved by this routine is almost certainly not gout The relief in acute gout, when it occurs, is dramatic and there can be no doubt of a positive response to the test.

4 Massage or physiotherapy, other than application of hot or cold compresses, is not indicated and may produce an exacerbation of the attack

5 Adrenocorticotrophic hormone (ACTH) relieves the acute attack (see Chapter 8 for a discussion of ACTH)

Interval management (between attacks)—Interval treatment is prescribed on the basis of the frequency and severity of attacks

1 Every patient who has had an attack of acute gouty arthritis should carry 1/100 gr colchicine tablets with him, just as the patient with angina carries nitroglycerin At the first sign of discomfort he should begin taking 1 tablet every two or three hours until relieved or until diarrhea occurs It is often possible by this simple means to abort or reduce the severity and duration of the gouty attack

2 Any patient subject to acute gouty arthritis who requires surgery should be especially prepared by use of a low purine diet and administration of salicylates and/or col-

therapeutic test (paragraph 3 below) is negative in rheumatic fever and positive in early gout attacks.

2. The palindromic type of rheumatoid arthritis is apt to behave clinically very much as gout. Sudden acute attacks in joints of the upper extremities with complete or almost complete recovery are apt to be atypical rheumatoid arthritis and, *in a woman, almost certainly so.* Such attacks occurring in the lower extremities of a man may be either gout or atypical rheumatoid arthritis. The skin over the joint involved by acute gout tends to discolor and to scale as the attack subsides, although this is not invariably true. One needs to be wary with this problem as the attacks of atypical rheumatoid arthritis, i.e., palindromic rheumatism, may be so brief that a false positive result of the colchicine therapy test may be recorded Sometimes a period of observation and trial treatment may be necessary to differentiate with certainty between atypical rheumatoid arthritis, i.e, palindromic rheumatism, and gout. Differentiation is especially difficult in the presence of acute intermittent arthritis of the *lower extremities in a man. Fortunately* one does not often meet this problem

3 Traumatic arthritis and osteoarthritis do not respond to the colchicine therapy test and should offer no serious problem in differential diagnosis (see the points in diagnosis of osteoarthritis, p 60).

TREATMENT

Treatment of the acute attack.—The routine to be followed during the attack is well established.

1. Absolute bed rest.

2. Protection of joints with cradle, footboard or cott batting

3 Colchicine (1/100 gr) every two to three hours, a tolerated, until diarrhea occurs. The diarrhea, if not self limiting, may be controlled quickly with paregoric The diarrhea, or therapeutic point, may develop within 24 hours but may be delayed as long as three days If nausea develops before the diarrhea, reduce the frequency of the colchicine administration Definite diarrhea (three or four watery stools) is the best guide to adequate dosage An acute attack of arthritis not relieved by this routine is almost certainly not gout The relief in acute gout, when it occurs, is dramatic and there can be no doubt of a positive response to the test

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Low Fat, Moderately Low Purine, High Carbohydrate Diet for Gout

Foods not Allowed		Foods Allowed
Sweetbreads		Lean beef
Anchovies		Lean lamb
Liver		Whitefish
Kidneys		Baked fowl
Brains		Eggs
Sardines		Mild cheese
Gravy		Fruits
Sauces		Vegetables other than those not allowed
Fat meat		White bread
Butter and fats (only as specified)		Skimmed milk
ALL spices and condiments		Sugar
Vegetables		Weak coffee
Spinach		Weak tea
Asparagus	Cauliflower	
Peas	Onions	
Dry beans	Mushrooms	

Sample Menu for Gout Diet*

Breakfast	Lunch	Dinner
1 serving of fruit 1 egg 1 slice white bread 1 teaspoon butter Weak coffee or tea, as desired	2 eggs or 2 oz mild cheese 2 servings of allowed vegetables 1 salad 1 serving fruit 1 slice white bread or 2 soda crackers 1 teaspoon butter or mayonnaise 1 glass skim milk or buttermilk Weak tea or coffee, as desired	1 serving (2 oz) lean, plain meat, fish or fowl, 3 days a week, other 4 days use 2 eggs or 2 oz mild cheese 2 servings of allowed vegetables 1 salad 1 serving fruit 1 slice white bread 1 teaspoon butter or mayonnaise 1 glass skim milk or buttermilk Weak tea or coffee, as desired

* Total calories should be adjusted to reduce weight if the patient is overweight

chicine for three to five days before surgery takes place

3. A patient subject to gouty arthritis should make every attempt to protect himself against undue trauma or injury Footwear and socks should be carefully selected and unnecessary stresses and strains to weight-bearing joints avoided

4. For patients having one mild attack or less yearly, the interval treatment is confined to routines 1, 2 and 3.

5 For patients having one or two severe attacks yearly, routines 1, 2 and 3 are followed and, in addition:

a) A low fat, moderately low purine, high carbohydrate diet

b) Aspirin, 60–80 gr daily for three days each week, because it increases uric acid excretion

c) If this routine does not provide satisfactory results, cinchophen may be substituted for aspirin, in doses of 7½ gr. three times a day for three days each week In gout, liver toxicity from cinchophen is extremely low or absent Nevertheless, patients taking cinchophen should be watched for evidence of toxicity (nausea, pruritus, jaundice, etc).

6 Patients who have repeated severe attacks yearly should follow routines 1, 2, 3, 5*a*, 5*b* and 5*c*, and add

a) Colchicine, 1/100 gr. three times daily for three days weekly, not on the same day that aspirin or cinchophen is taken

CHAPTER 7

Collagen Diseases

THE COLLAGEN DISEASES, strictly speaking, include all diseases in which there is characteristic fibrinoid degeneration of collagen. Collagen is a product of fibroblasts. Fibrinoid degeneration consists of swelling of the mucoid ground substance, together with proliferation, degeneration and necrosis of fibroblasts. Inasmuch as these changes occur in connective tissue, diffuse collagen disease may involve many different areas of the body. Just as fibrinoid degeneration is the common denominator of these diseases pathologically, *multiple system involvement* is the signboard pointing to the clinical diagnosis.

The following disease syndromes are usually classified as diffuse collagen diseases.

1. *Periarteritis nodosa (polyarteritis, necrotizing vasculitis)*—The fibrinoid degeneration in this disease tends to localize in the media and intima of the medium-sized and small arteries The lumen of the artery may be obliterated and thrombosis or hemorrhage may occur. The blood vessels of the heart, kidneys, brain or any other part of the body may be involved. Clinically the disease resembles a severe progressive infection, with fever, weakness, loss of weight,

muscle and joint pains and, usually, an intermittent downhill course Laboratory studies reveal a rapid sedimentation rate, usually albumin and red cells in the urine, often eosinophilia with leukocytosis. The diagnosis may often be established by muscle biopsy. *Multiple system involvement,* including joints, kidneys, heart, etc., should suggest the diagnosis

2 *Lupus erythematosus disseminatus* —This too is a disease of connective tissue due to collagen necrosis. The necrosis may be very widespread, involving blood vessels, pleura, serous membranes, central nervous system, heart, kidneys, skin, lymph nodes, etc When blood vessels are affected there are usually endothelial proliferation and occlusion, producing the same end-result as periarteritis nodosa. The name is misleading, because the skin lesions play a very minor role and often are not present. Clinically it is characterized by fever, weight loss, fatigue, pleurisy, muscle and joint pains, gastrointestinal disturbances and, occasionally, the typical butterfly lesion across the bridge of the nose and cheek. Pleuropericardial and pericardial effusions are common. Laboratory studies usually reveal leukopenia, albuminuria and sometimes hematuria The sedimentation rate is increased The diagnosis can usually be made by muscle biopsy and by sternal marrow examination *Multiple system disease,* usually involving joints, serous membranes, kidneys and skin, should suggest the diagnosis

3 *Dermatomyositis* —This necrotizing collagen disease chiefly involves the skin, skeletal muscles and the heart There is seldom direct blood vessel necrosis, although the vessels are involved secondarily through disease of the surrounding structures The clinical course is characterized by muscular contractions and fibrosis of the skin and fascia,

producing fixation of joints, but the most striking feature is the muscular weakness, which is progressive, usually with pronounced tenderness on pressure of the muscles. Late in the disease the muscles become firm, indurated and board-like. Laboratory studies are seldom helpful. The diagnosis usually may be made by muscle biopsy.

4. *Scleroderma* involves chiefly the skin, gastrointestinal tract, heart and other viscera. Muscle and connective tissue are replaced by an overgrowth of fibrous tissue. Induration and constriction are common. Circulation is impaired. There are muscle and joint pains and the constriction of fibrous tissue may limit joint motion. Pulmonary and myocardial fibrosis may occur The muscular layers of the gastrointestinal tract may be replaced by fibrous tissue. The course usually is slow and may be spontaneously arrested at any time If the disease progresses, weakness and emaciation increase and death usually is due to secondary infection. X-ray examination frequently reveals calcifications, usually situated deep in the tissue around the large joints. Gastrointestinal x-rays may show strictures or irregular dilatations, characteristic of the disease. Laboratory studies are not helpful Once the disease is established, the diagnosis is usually obvious.

Rheumatoid arthritis, rheumatic fever and serum sickness are sometimes grouped pathologically with the collagen diseases, although they are not included in this discussion.

DIAGNOSIS

As will be readily seen, the collagen diseases, though called by different names, often involve the same structures

and present many changes in common. Indeed, it is not always possible to give an exact name to many of the cases seen However, it should be possible to make the diagnosis of diffuse collagen disease if it is always kept in mind as a possibility in every patient seen with suspected rheumatic disease

Diffuse collagen disease may imitate many other diseases, but in our experience it is most frequently confused with rheumatoid arthritis and rheumatic fever. Rheumatoid arthritis is also a systemic disease but almost never presents major clinical evidence of *multiple system involvement.* The differential diagnosis is therefore a quantitative one, and the more extensive the clinical and laboratory evidence of multiple system involvement, the more probable is the diagnosis of diffuse collagen disease

The following case histories indicate many of the important diagnostic points and also illustrate some of the difficulties that are likely to be encountered in making an early diagnosis.

REPORTS OF CASES

CASE 1 —A woman (V K), aged 45, suffered from polyarthritis which was diagnosed as rheumatoid arthritis She had been well until two years before her death when she noticed a rather insidious onset of painful swelling with some local heat in both knees Subsequently the wrists, elbows and shoulders were painful and the proximal interphalangeal joints and terminal interphalangeal joints of both hands were aching and stiff The patient lost about 20 lb. in two months The sedimentation rate was increased to 88 mm in one hour by the Westergren method. The white blood cell count at the onset of the illness was 4,000, with a marked shift to the left There was moderate hypochromic,

microcytic anemia. The patient noticed some weather effect and she received a moderate amount of relief from aspirin The diagnosis was rheumatoid arthritis. She was given vitamins and told to apply heat to the joints and later was advised to come to Tucson.

Examination about one year before death disclosed swollen painful knees with some limitation of motion on the right. The proximal interphalangeal joints of both hands were tender and somewhat painful but there was no evidence of swelling or thickening The shoulders were moderately limited in abduction, with pain at the extremes. The patient was thin, undernourished, the blood pressure was 92/60, the pulse 110 and the rhythm regular The positive points on physical examination were essentially inanition and the polyarthritis The white blood cell count was 3,200, with a pronounced toxic blood picture, a shift to the left and toxic granules in the polymorphonuclear leukocytes. The sedimentation rate was 120 mm. in one hour by the Westergren method. There were 6-8 red blood cells per high power field in the urine and 2 plus albuminuria. The electrocardiogram revealed nonspecific changes consistent with a diffuse type of myocarditis. The chest x-ray was essentially unremarkable. A muscle biopsy revealed changes compatible with lupus erythematosus disseminatus

The subsequent course was progressively severe. The temperature began to rise to as high as 103 F. in the late afternoon. The white blood cell count fell to a total of 2,000 and the hemoglobin content to 58 per cent. The urinary changes became more severe and cardiac arrhythmia developed. There was at one time a moderate pleural effusion on the right, and there were two distinct episodes of aphasia, dizziness and weakness of the right arm and leg. Careful examination of the eyegrounds at intervals revealed a few flame-shaped hemorrhages and one or two small arterioles that seemed to be obstructed. Moderate retinal edema developed The blood urea nitrogen value slowly began to rise. Multiple areas of tenderness in various muscles and tendon sheath swelling appeared and the size of the swollen joints increased.

The patient's weight continued to decline, nausea and vomiting became a feature and suddenly there developed complete obstruction of the left brachial artery This was followed by gangrene of the arm, requiring amputation. The patient died two months later, and at autopsy the diagnosis of lupus erythematosus disseminatus was proved.

At no time did this patient exhibit skin lesions of the typical butterfly type over the bridge of the nose and the cheek There were no alterations in the skin other than those secondary to the nutritional disturbances and the patient's illness The suggestion that the disease was not rheumatoid arthritis but lupus erythematosus disseminatus arose out of the finding of leukopenia, alterations in the urine consistent with a diffuse type of vascular nephritis and the involvement of the heart muscle Subsequent involvement of the brain, of the pleura and of the gastrointestinal tract further confirmed the diagnosis. The case is a typical example of this fatal disease, although the occlusion of such a large artery as the brachial is somewhat unusual (Fig 73)

Case 2—A young woman (M V), aged 19, had a disease which was diagnosed as early rheumatoid arthritis for one month before the appearance of leukopenia and diffuse arteritis She died one year after the onset of her illness and at autopsy the clinical diagnosis of lupus erythematosus disseminatus was confirmed (Fig 74)

Case 3—A woman (V G), aged 57, had a disease which was diagnosed as rheumatoid arthritis for five years before the onset of *multiple system involvement* The patient had profound weakness, exquisite tenderness of the muscles and diffuse systemic involvement Biopsy revealed cutaneous ulceration and disturbances in the muscles characteristic of dermatomyositis. She died six years after the onset of the illness, and at autopsy the diagnosis of dermatomyositis was confirmed

Case 4—A Mexican man (J O), aged 33, was said to have glomerulonephritis No articular disease was present Subsequently the diffuse picture became clear and, on histologic examination, polyarteritis was evident At autopsy eight months



Fig 73 (top) —Lupus erythematosus disseminatus; Case 1. Transverse section of a thrombosed brachial artery of the left upper arm The intima

power photomicrograph of an arteriole, showing intimal disintegration and vacuolation of the muscular cells plus inflammatory changes in the adventitial coat Many nuclei are undergoing karyorrhexis, and the vessel shows all stages of inflammation and degenerative attempts at repair.

after the onset of the illness the diagnosis of polyarteritis was confirmed

CASE 5.—A woman (E R), aged 45, had a disease which was considered to be rheumatoid arthritis for one year before the clinical and pathologic diagnosis of lupus erythematosus disseminatus was made At this time, leukopenia developed and the histopathologic changes of lupus erythematosus disseminatus were noted She was given Benadryl in doses of 300–500 mg in 24 hours and remained on this medication She went back to work as a school teacher and at the time of writing had enjoyed an asymptomatic course for over a year (Fig. 75)

CASE 6.—A man (G K), aged 45, had a disease which had been diagnosed as rheumatoid arthritis for 11 years The story of the early illness was characteristic of gouty arthritis, with sudden attacks of exquisitely painful arthritis in the peripheral joints followed by complete subsidence of pain Subsequently, however, chronic polyarthritis and tenosynovitis developed, and three years before his death there was evidence of *multiple system involvement* A muscle biopsy revealed characteristic changes of polyarteritis. Twelve years after the onset of the attacks of arthritis the patient died At autopsy, polyarteritis was found (Fig 76)

CASE 7.—A man (W.H), aged 55, was told he had rheumatic fever nine months before the detection of *multiple system involvement* Histologic examination confirmed the diagnosis of polyarteritis. He was given Benadryl in doses of 300-500 mg in 24 hours, following which his course was one of progressive improvement At the time of writing, one year after the final diagnosis was made, he was almost symptom free

CASE 8.—A woman (V G), aged 30, had a disease which was diagnosed as rheumatic fever for one month before the detection of lupus erythematosus disseminatus Histologic examination confirmed the diagnosis This patient, although she had a stormy course, was at the time of writing considerably improved She was afebrile, up and about and had no complaints She too was given Benadryl in doses of 300-500 mg per 24 hours and con-



FIG 75 —Lupus erythematosus disseminatus, Case 5 Characteristic distribution of skin lesions on the hands and shoulders The face was not typically involved



FIG 76 —Polyarteritis, Case 6. Associated changes in this patient included Dupuytren's contractures, especially in the left hand, and darkened areas of the palms and finger-tips—so-called "liver palms."

tinued with varying amounts of the drug during the succeeding three years without recurrence of the disease

CASE 9 — A woman (F.A.), aged 30, had had asthma for eight years One year before her death the condition was diagnosed as rheumatoid arthritis and she was treated with vaccines. Finally she was referred to Tucson and six months before death there was evidence of polyarteritis Diagnosis was confirmed by histologic examination. Gradually, symptoms of *multiple system disease* developed and the patient died At autopsy the diagnosis of polyarteritis was confirmed (Fig 77)

CASE 10 — A woman (E R), aged 40, had a disease which was diagnosed as rheumatoid arthritis for 13 years She had been treated with gold, transfusions, vaccines and vitamins and had had a variable course. When she was first seen by us, three years before her death, the condition was typical of *rheumatoid* arthritis. Within a year, however, leukopenia developed, and on careful study *multiple system involvement* was evident Finally the classic lesions of lupus erythematosus appeared on the face, and at autopsy the diagnosis of disseminated lupus erythematosus was confirmed

CASE 11 — A man (R W.), aged 32, had a disease which was diagnosed as rheumatoid spondylitis and peripheral rheumatoid arthritis for eight years Five years after onset of his joint complaints he came to Tucson, and roentgenographic studies confirmed the diagnosis of rheumatoid spondylitis However, within six months of that time and two years before his death, leukopenia gradually appeared, and subsequently lupus erythematosus and *multiple system involvement* became evident The clinical course was variable, and although the patient took 600 mg of Benadryl per day, he eventually died At autopsy the morphologic changes of lupus erythematosus disseminatus were observed (Fig. 78)

Differential diagnosis —Some of our patients may have had coincidental rheumatoid arthritis, and it is interesting to speculate that perhaps the rheumatoid arthritis gave way to

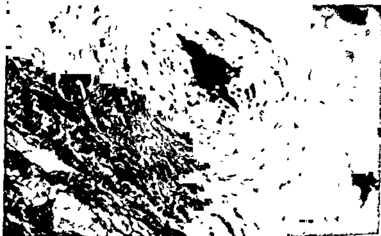


Fig 77 —Polyarteritis, Case 9 Photomicrograph showing a collar of inflammatory cells around an arteriole, vacuolation of the subintimal layer, in the adventitia, a tremendous laying down of collagen material, and a liberal sprinkling of acute inflammatory cells throughout



Fig 78 —Lupus erythematosus disseminatus, Case 11. Section from a muscle biopsy shows a nest of inflammatory cells among the muscle fibers, which stained poorly and inconstantly The transverse striations of skeletal muscle have disintegrated into a homogeneous mass in several muscle bundles

the different clinical and morphologic picture of collagen disease In any event, it is of some interest that of our 11 consecutive patients, nine suffered for a considerable period of time from what appeared to the referring physician to be a form of rheumatic disease It would thus seem that when we are faced with an articular problem we must be exceedingly cautious in concluding that the patient suffers from either rheumatoid arthritis or rheumatic fever and that the differential diagnosis between a collagen disease and any rheumatic disease in the early stages may be exceedingly difficult

Of considerable help in the differential diagnosis is the appearance of, the total number of and the kind of white blood cells found on routine blood examinations In a considerable proportion of cases of collagen disease, leukopenia is a part of the picture and may offer a helpful guide Bone marrow studies are often characteristic More important, however, are careful history and physical examination, which may reveal appreciable changes in the kidneys, the heart and the peripheral vessels, i e , *multiple system involvement,* all of which should suggest collagen disease. It has been well demonstrated that patients with rheumatoid arthritis may have some cardiovascular changes seemingly associated with peripheral arthritis; however, in our experience, these cardiovascular changes are never as profound as those which are seen in collagen disease

Even though the patient has in the past presented a typical picture of rheumatoid arthritis or rheumatic fever, any change suggesting *multiple system involvement* may mean the initiation of a new disease or an error in the previous diagnosis It appears that the diagnosis in six of

our 11 cases was erroneous, and it seems probable that four patients either had insensible progression of the rheumatic disease into collagen disease or developed an independent new disease. In one (Case 4), glomerulonephritis was the first manifestation of the *multiple system disease*. It seems likely that *there is a common denominator* for collagen disease and rheumatic disease in the processes of intrinsic metabolism; and experience by us and others with cortisone and ACTH lends strong support to this concept. Collagen disease should be considered in any patient with an articular disturbance who shows evidence of *multiple system involvement*. It should be included in the differential diagnosis of any arthritis and especially should be thought of in patients with typical rheumatoid arthritis or rheumatic fever who subsequently have disturbances or changes in organs not ordinarily affected by a rheumatic process.

In summary, think of collagen disease as a possibility when any patient with joint complaints has any one of the following symptoms:

1. Leukopenia.
2. Hematuria or albuminuria.
3. Pleural effusion or pneumonitis.
4. Myocardial changes.
5. A skin rash or skin changes of unusual character, i.e., lupus (Fig. 75) or scleroderma (Fig. 79).
6. Central nervous system alterations.
7. Any unexplained systemic disorder.

If diffuse collagen disease is a possibility, muscle biopsy should be done. A positive result may clinch the diagnosis

But a negative result does not rule out a diagnosis of collagen disease

TREATMENT

Rich was able to produce rheumatoid lesions in rabbits by the injection of horse serum Subsequent experimental work has demonstrated that the effect of horse serum noted by Rich can be blocked by antihistamine medication. We have

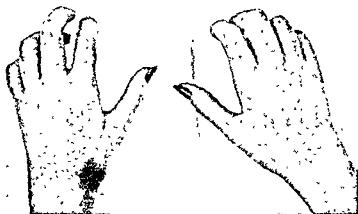


FIG 79 —Scleroderma

been able to treat successfully three of four patients having collagen disease by administration of an antihistamine (Benadryl) in large doses (300–600 mg per 24 hours) In these three patients the condition was diagnosed in a relatively early stage. Recognizing the variable course of collagen disease, we believe nevertheless that Benadryl may have some rational place in the treatment of the usually hopeless con-

dition, since the three patients are alive and apparently symptom free. Other agents—bismuth, gold, transfusions and a wide variety of other substances—have been unsuccessful in the treatment of these unfortunate patients, and thus far there is no specific cure for collagen disease.

CHAPTER 8

Cortisone and Adrenocorticotrophic Hormone (ACTH)

PROBABLY NEVER IN the history of medicine have so much information and misinformation been given the public by magazines, newspapers and radio as has been the case since the first reports on the "miraculous results" and later of the "terrible hazards" of these two substances. It was in April, 1949, *that Hench, Kendall, Slocumb and Polley, of the Mayo* Clinic, made the first public announcement that these two substances would produce a dramatic remission in rheumatoid arthritis.

Cortisone is one of the many steroids said to be produced by the adrenal cortex. Its structural formula is known. Its chemical name is 17-hydroxy-11-dehydrocorticosterone. It is at present prepared by partial synthesis from ox bile by Merck & Co. and supplied as the acetate salt in suspension (25 mg. per cc.). The supply from this source is limited and expensive. Through the recent improvement of manufacturing processes the yield has been increased and the cost reduced. Allocation of cortisone for research was, until January 1, 1950, controlled by a committee of the National Re-

search Council. In 1950, Merck & Co., with an advisory board, are making allotments to research workers at a token price of $150 per gram and hope to increase the supply at a lower price. Many other pharmaceutical companies, as well as Merck & Co., are seeking other sources of cortisone soy beans, yams and certain African plants are being investigated. Clinical trials with other products are not sufficient to permit evaluation. The chemists are also busy attempting complete synthesis of cortisone and similar substances. The future available supply may be markedly increased by success in any of these efforts.

ACTH (adrenocorticotrophic hormone) is one of the hormones produced by the anterior lobe of the pituitary. Its function is to regulate the activity of the adrenal cortex It is at present prepared largely from the pituitaries of hogs. The Armour Laboratories have pioneered the preparation of this substance. It is packaged in ampules of 10, 25 and 40 mg. (Armour Standard). Obviously, again, the supply from this source is extremely limited and expensive. Armour and Company and other chemists and pharmaceutical companies are seeking cheaper and better supplies from natural and synthetic sources. The allocation of ACTH for research has been carried out by Armour and Company through Dr. John Mote, Medical Director, and an advisory research board. *All statements in the following discussion on ACTH refer only to the Armour Standard; we have not had experience with ACTH from other sources.*

Between April, 1949, and April, 1950, we have administered cortisone to 42 patients and ACTH to 50 patients with rheumatoid arthritis. Sixty of them have been observed in the hospital on metabolic control, and numerous

laboratory and chemical estimations have been made We have been chiefly concerned with uncovering knowledge regarding *the mechanism of remission* and in determining, if possible, the essential chemical changes which are responsible for the remission when cortisone or ACTH is administered This experience, together with observations of other careful investigators, forms the basis for the discussion to follow The chemical change essential for the production of remission has not been identified with certainty, but much knowledge has been gained regarding the metabolic changes that occur during administration of these substances Nitrogen, potassium, phosphorus, uric acid and salt and water balances are affected. Globulin-albumin ratios, sedimentation rates, the number of circulating eosinophils and the blood levels and urine excretion of amino acids are altered No conclusive evidence has been offered that any of the now measurable changes are essential or specific to the remission factor. The discovery of the secret of the essential remission factor may require much additional research, but the use of these two substances as research tools makes eventual success almost a certainty

EFFECTS IN VARIOUS DISEASES

Cortisone will (1) produce a temporary remission in the activity of rheumatoid arthritis, (2) produce a remission or markedly improve the collagen diseases, such as lupus erythematosus disseminatus, periarteritis nodosa and dermatomyositis, (3) dramatically suppress, if not halt, rheumatic fever, though in continuous cyclic rheumatic fever it may only produce a temporary remission

Cortisone cannot: (1) restore destroyed joints, (2) correct deformities with joint damage of any duration; (3) be effective unless given continuously or intermittently in rheumatoid arthritis, the disease recurring sooner or later after cortisone is discontinued; (4) be given continuously over a long period without danger of toxic effects.

ACTH will: (1) produce a temporary remission in the activity of rheumatoid arthritis; (2) abruptly halt the course of rheumatic fever and, unless the disease is recurrent, will terminate the attack; (3) produce dramatic response in an acute attack of gout and possibly make the patient more susceptible to the favorable effects of colchicine; (4) produce at least temporary remissions in the collagen diseases named previously.

In addition to the foregoing so-called rheumatic diseases, ACTH has had wide research trial in many other disease syndromes. It will clear rather rapidly such allergic skin disturbances as hives, penicillin reactions, gold dermatitis and exfoliative dermatitis. Psoriasis is improved or cleared. Virus pneumonia, infectious hepatitis and lobar pneumococcic pneumonia are reported to be favorably influenced Long remissions have been described following administration of ACTH in myasthenia gravis Asthma, ulcerative colitis and some eye inflammations respond well, sometimes dramatically.

ACTH cannot: (1) restore destroyed joints; (2) correct deformities with joint damage of any duration, (3) be effective in rheumatoid arthritis unless given continuously or intermittently, because the disease recurs sooner or later after ACTH is discontinued; (4) be given with certainty over a prolonged continuous period without hazard of toxi effects.

Research studies have indicated that ACTH is not effective in such diseases as poliomyelitis, tuberculosis, multiple sclerosis, Parkinson's disease and psychoses.

DOSAGE

Cortisone.—Three hundred (300) mg on the initial day of treatment in two divided doses, and 100 mg in a single dose daily thereafter has been the standard initial procedure. The duration of treatment in our series has varied from eight to 14 days. One patient of the 42 required 200 mg daily to produce remission. Usually the period of eight to 14 days is adequate to achieve marked improvement.

ACTH.—This hormone is extremely active and in large doses is capable of producing profound and disturbing metabolic changes. It is therefore desirable to use the minimal effective dose. Of 45 patients with active rheumatoid arthritis who have received 40 mg or less per 24 hours (10 mg every six hours), only one has required larger doses. The standard routine of dosage that we have adopted is 10 mg every six hours, to 40 mg per day, for 10–20 days. This is sufficient usually to produce striking improvement.

UNDESIRABLE PHYSIOLOGIC EFFECTS
(Toxic Reactions)

Following the aforementioned dosage schedules, three patients on cortisone developed very slight rounding of the face which disappeared within two weeks after the medication was discontinued. One patient, who had previously had a coronary occlusion, had mild pulmonary edema on the seventh day of cortisone administration. Of five patients receiving 80–160 mg of ACTH daily, one had transient hyper-

tension and one, edema. Of the 45 patients given 40 mg. of ACTH or less daily, three had transient rounding of the face, which disappeared within a few days. On the 40 mg. daily schedule, there were no other measurable disturbances such as hypertension, glycosuria, hyperglycemia, sodium and water retention, hirsutism, mania or voice changes. Exhilaration and mild euphoria with some insomnia were occasionally encountered both with cortisone and with ACTH.

CLINICAL RESPONSE

The clinical response in patients with active rheumatoid arthritis to the administration of cortisone and ACTH is, so far as we can tell, identical Within 48 hours the patients described a feeling of well-being and a decrease in joint stiffness, rest pain, motion pain and joint tenderness. There was rapid recovery of strength Nearly all patients show a pronounced increase in appetite and energy. There was marked improvement in strength as measured by the grip. Joint swelling began to diminish by the third day. Patients in the early stages of the disease, with little or no joint destruction, usually underwent complete clinical remissions. Figures 80 and 81 illustrate the clinical course.

All 92 patients exhibited measurable remission Eighteen (with minimal joint destruction) experienced a complete clinical remission; 70 had striking improvement. Four evidenced measurable remission of some degree but were unsatisfactory for further studies.

Duration of improvement.—The period of improvement has varied in this group of patients from one day to more than eight months. The average patient begins to relapse the

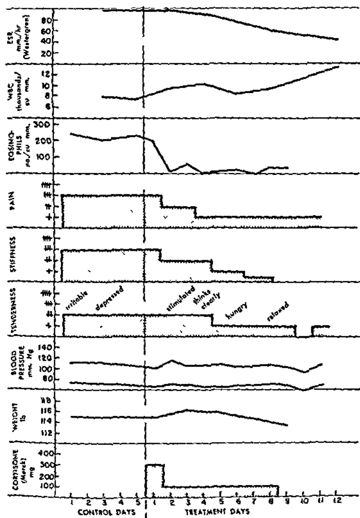


FIG 80 — Typical response of rheumatoid arthritis to cortisone

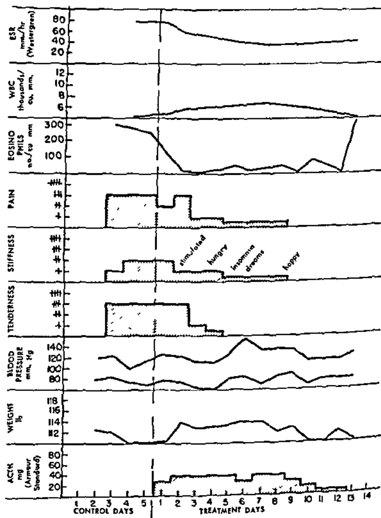


FIG. 81 —Typical response of rheumatoid arthritis to ACTH

second week and gradually within a month returns to his previous state. Three patients have maintained 75 per cent of improvement for more than six months Fifteen patients maintained definite improvement for more than three months Three relapsed within 48 hours The duration of remission is unpredictable The rest of our group are being studied on maintenance routines so are not available for a report on duration of remission.

Attempts to maintain remission.—When a patient has achieved a marked improvement with cortisone or ACTH, on the dosage schedule described, the physician is faced with the problem of attempting to maintain this remission Two general methods are open to him He may discontinue the medication and depend on repeat courses in the hope that the remission will be maintained longer after the additional courses, or he may elect to try the smallest effective maintenance dose given continuously

1. ACTH Remission can, in some instances, be maintained by continuous small doses One patient has been maintained in complete remission for more than six months on 5–10 mg daily. Ten patients have retained maximal improvement on 10 mg daily, in divided doses, for three to six months In two of these, transient edema developed Interrupted courses of treatment have been tried on seven patients who have experienced a maximal remission on four occasions during the past year on 20 mg or less daily (divided doses), for a treatment period varying from 10 to 14 days It is our impression that divided doses—a minimum of twice daily and preferably four times daily—are superior to the single dose

2. Cortisone Remissions in some instances have been

maintained by others with doses of 50–75 mg. daily. One hundred (100) mg. three times weekly has been tried. We have limited our studies to the interrupted course method, using the dosage schedule already described and allowing a three week rest after completion of the course before repeating the course of cortisone It is not known whether improvement will be prolonged after discontinuance of the medication We have observed no serious toxic effects so far in the 30 patients on this routine.

3. The basic treatment program, consisting of corrective and postural exercises, correction of anemia, blood transfusion, etc., has been helpful in maintaining muscle tone and joint motion gained during the ACTH or cortisone remission

4. Miscellaneous Gold therapy in conjunction with ACTH or cortisone is being tried, but insufficient time has passed to allow judgment of its advantages Many medications, including some of the other steroids, have been used in an attempt to maintain the remissions, but so far without uniform benefit

PRECAUTIONS IN USE OF ACTH AND CORTISONE

Although it is true that patients receiving low doses of ACTH and cortisone (in the dosage described for a limited period) seldom have undesirable physiologic effects, the most careful observations may be required to avoid these complications Rounding of the face, sudden weight gain, edema, severe fatigue, elevation of blood pressure, glycosuria or hyperglycemia and disturbing mental changes may occur Sodium and sodium chloride intake should be reduced to a minimum if it is necessary to give larger doses or to continue

treatment for longer periods This should be done also whenever there is evidence of water or salt retention Fluid limitation may also be helpful Potassium excretion in the urine is increased with the administration of ACTH, the symptoms of potassium deficiency, such as weakness and characteristic electrocardiographic changes, can be eliminated by regular administration of one of the potassium salts in doses of 3–5 Gm daily

ACTH and cortisone have a profound effect on carbohydrate metabolism It is probable that in the dosages discussed neither hyperglycemia nor glycosuria will occur in the nondiabetic individual These substances also increase the insulin requirement in the diabetic, and patients known to have diabetes should not be given ACTH or cortisone unless the most rigid conditions of study are carried out The evidence points to the probability that glycosuria and hyperglycemia that have developed during a study period have disappeared after a week or two Patients with chronic nephritis should in general be excluded from ACTH and cortisone therapy Patients with psychopathic personalities, known psychotics or patients with a known history of psychosis should not receive ACTH or cortisone owing to the known effects of these substances on cerebral function

ACTH EFFECTS ON COMPLICATING DISEASES

In addition to rheumatoid arthritis, one of the following conditions in one or more patients were dramatically relieved by ACTH asthma, Sjogren's syndrome (keratoconjunctivitis sicca, with dry mucous membranes), psoriasis, severe penicillin sensitivity reaction, atopic eczema, virus hepatitis with

jaundice, hyperthyroidism, gold dermatitis and various inflammations of the eye, including severe iritis.

COMMENT

It is obvious from the foregoing data and information that much more research work must be done before these substances are suitable or available for routine treatment of rheumatoid arthritis. More information must be gained regarding long-term administration, ways must be found to prolong remissions once they are achieved, new substances must be sought that are cheaper and more easily available and, above all, the remission factor X should be sought It it quite conceivable that when the specific mechanism of remission is known, it may be produced and maintained at the cost of a few cents instead of many hundreds or thousands of dollars.

CHAPTER 9

Prevention and Correction of Deformities

PREVENTION OF DEFORMITY

RHEUMATOID ARTHRITIS

DRUGS AND COMPOUNDS, including cortisone and ACTH, will not mobilize a frozen joint nor make the deformed cripple jump from his wheel chair and dance The new compounds hold great promise as guides to discovery of a way to stop the inflammation and progress of active arthritis, but they will not restore damaged joints nor release tight adhesions With the expectation of a new method to stop the progressive destructive activity of arthritis, it becomes more than ever before the duty of every physician treating arthritis to teach every patient methods of preventing deformity and maintaining function

Not only can many of the deformities be prevented, but joint function can be maintained by simple measures that every patient can follow By following a good program, the patient has a chance to salvage a useful body when the disease process either burns out or is arrested medically, be it after six months, six years or longer.

Prevention of deformity must start at the onset of the disease with a careful explanation to the patient of the nature of the disease and its serious consequences. If the onset is acute and sufficiently severe, the patient is usually quite willing to accept a full routine of treatment to get relief. With the more common slower, insidious onset that characterizes so many cases, it is more difficult to convince the patient that he must follow a program that will often seem drastic or out of proportion to the amount of discomfort he is suffering The significance of the early swollen finger joint or wrist is often minimized and the patient goes on straining the joint in his daily occupation. The first signs of rheumatoid arthritis which appear in the feet are frequently mistaken for fallen arches and the patient may walk on them unadvisedly for months. It is during the early months of this disease, before too much damage has been done, that the patient should have the benefit of a careful explanation of the problem At this point, if the physician will take the time for a careful evaluation and explanation, much crippling damage can be prevented. Not only will he help his patient but he will avoid the need for many hours of explanation later when the patient is dissatisfied and is still looking for an easy cure

The first thing the patient must understand is that rheumatoid arthritis is a progressive crippling disease and that if it goes on unchecked for many years, as it does in most cases, it can leave him a helpless cripple or, at least, seriously handicapped. The patient who understands the insidious nature of the disease will much more readily accept a broad program that is going to protect him for years to come Until we have found a short cut, a way to stop the activity and the

progress of the disease, the patient must learn and follow a basic program to protect his joints and maintain motion, regardless of how long the disease may last. He should also understand that the disease involves all parts of the skeletal system—demineralization of bones, erosion of cartilage, atrophy of muscles, inflammation of joint capsules, weakening of ligaments and tendons around joints, all of which cause progressive stiffening, loss of motion and ultimate ankylosis unless active preventive steps are taken.

After showing the worst of the picture to the patient, one can encourage him by explaining that there are ways of minimizing deformity and maintaining a large measure of joint function. To accomplish this objective, the patient must fully understand and be carefully directed in the execution of the three fundamental parts of his program: (1) avoidance of strain to joints and muscles, (2) proper rest, and (3) proper exercises

1. Avoidance of Strain to Joints and Muscles

Many cripples have been made by the very bad advice that they must "keep going" because if they ever quit they will not be able to walk again Consequently, the patient often struggles along on sore feet, ankles, knees and hips, increasing the joint damage and deformity with every step until he can go no longer This bad practice has been generally followed simply because no one has told the patient that he could better preserve motion and strength by exercising in bed without strain on his joints. Some types of occupation are often helpful in maintaining strength and motion, but too often we find the occupation harmful, for example,

duties of the housewife may cause daily trauma or strain to the affected joints. The individual may be able to continue work for an indefinite period, but he runs the risk of progressive and total destruction which may completely disable him ultimately. If the patient is lucky, the disease will burn out before he becomes too badly crippled; unfortunately, these cases are in the minority.

It is here that it becomes important for the doctor to evaluate the extent and progress of the disease. The problem is to make the joint function last as long as the patient needs it for economic reasons and for his happiness. The patient should be encouraged to work as long as his occupation is not damaging. Some patients with minimal involvement may be allowed or encouraged to do full-time work, others part-time work, and some may have to change the type of occupation The patient with a severe form of arthritis ultimately may be much better off economically to take six months off from work at the onset rather than to go on until he becomes crippled for the rest of his life. To the young patient, wage earner or housewife, the doctor has an exceedingly serious problem in the advice he is going to give. In outlining a program of treatment, many factors have to be considered—the extent and severity of the disease, rapidity of its progress, estimation as to prognosis and the individual's temperament, economic status, etc.

When there is active inflammation in the weight-bearing joints, the patient should stay off his feet as much as possible If the inflammation is minimal and his posture not too bad, he may be allowed to walk short distances to the bathroom, table or automobile, provided this does not aggravate the joint disturbance or the deformity. *Pain during use or exer-*

cise usually has no significance so long as it does not persist after exercise and is not any worse when the same amount is done the next day. This "rule of tolerance" is an important guide and more is said about it later (p 144).

When there is a deformity in the foot or ankle or a flexion contraction of the knee or hip, there is naturally a strain on



Fig 82.—Size, shape and location of the foot support are important in rheumatoid arthritis

the joint on weight-bearing This strain, through poor posture and mechanics, occurs often enough to maintain signs of activity in the joint, such as heat, redness, swelling and soreness This fact can be demonstrated by disappearance of signs of active arthritis when a deformity is corrected and proper weight-bearing posture established.

Feet.—Feet are commonly involved at the onset of rheu-

matoid arthritis. In the early stages, before x-ray changes develop, it is sometimes difficult to differentiate between the pain of arthritis and the pain of a fallen arch. If there is active arthritis in the feet that cannot be relieved by the wearing of proper shoes and supports, the patient should not be al-



Fig 83 (top).—A good sturdy shoe with metatarsal bar is essential to prevention of arthritic deformity.
Fig 84 (bottom).—The arthritic patient is not permitted to wear slippers

lowed to walk or at least should walk only minimal distances. When the pain and symptoms can be relieved by proper shoes, supports, metatarsal pads, bars, etc., the patient may walk to tolerance.

Proper support for the foot is indicated to give correct



FIGS 85 AND 86 *(top)*—Tension in the arthritic foot is satisfactorily regulated by use of elastic bandages Elasticity is restored to the bandage by washing every few days

FIG 87 *(bottom)*—Example of a footboard that can be devised to prevent footdrop

balance and posture to relieve strain to the joints. It is usually sufficient to prescribe a low-heeled, straight-last, strong shoe, of adequate size, to which can be added sponge-rubber longitudinal arch supports and metatarsal pads as required (Figs 82 and 83). Soft slippers aggravate sore feet and increase deformity (Fig. 84). High narrow heels may produce strain on all weight-bearing joints as well as throw too much weight on sore metatarsal joints. The longitudinal and metatarsal pads must be of the correct size, large enough and placed properly in the shoe Adjustment of position and change to new ones is often necessary. Metatarsal bars placed on the outside of the shoe are often an additional advantage and remove pressure from sore metatarsal heads Wedging of the shoe may be necessary to give proper alinement (see p. 169). For further relief of strain on the longitudinal arch and ankle, a small 2 in. elastic bandage is helpful (Figs. 85 and 86). Dependent edema is very common when circulation is poor and the patient sits too long. Edema is increased by garters, so their use is forbidden. The patient should never allow the edema to persist and, when possible, the legs are elevated long enough each day to eliminate swelling If the patient must be on his feet it may be necessary to use elastic bandages or stockings Relief of strain on the foot and ankle at rest is also important. For the patient in bed, a footboard (Fig 87) is suggested to prevent footdrop Medium-sized sandbags can be placed on the outside of the foot and leg on the bed to prevent external rotation and eversion (Fig 87).

Knees.—If treated adequately and early, 100 per cent of the flexion contraction of knees can be prevented With knee involvement the patient seeks relief in flexion Pillows under



FIG 88 *(top)*—A means of relief commonly assumed by patients with arthritis in the knees This contributes greatly to flexion contraction and deformity

FIG 89 *(bottom)*—For the patient with arthritis of the knees, ordinary boards should be placed full length under the mattress Blocks elevate the bed to lessen strain on the joints when getting in and out of bed

the knees, which give relief, simply add to the flexion contraction and have been the greatest curse in producing deformity (Fig 88) With any more than the very minimal activity of arthritis in the knees, the patient should not be allowed to bear weight. Walking with inflamed knees, even though they are still straight, may increase the inflammation and the active arthritic process With a bent knee th[illegible] vation is prop[illegible]

fore wei[illegible]

on[illegible]

fir[illegible]

e [illegible]

[illegible] and sandbags hold the knees in the correct position for maximal rest and relaxation. Without the sandbags the feet tend to evert, causing torsion and chronic strain at the knees even at bed rest (Figs 90 and 91).

If there is enough "give" to the bed so that the knee cannot be rested in maximal extension, a small towel is folded and placed under the ankle to insure full extension (Fig 92).

PLASTER SHELLS (CASTS).—*If inflammation and pain in the knee are not relieved by rest on a flat bed, a plaster shell is recommended* When the knee is acutely inflamed it may be necessary to give a narcotic or even a light anesthetic to apply the first cast To make a rest cast for the knee, the leg is wrapped in sheet wadding from the hip down over the foot and well above the toes, using two or three layers. After this, 6 in rolls of plaster of paris bandage are used to make a solid leg cast extending from the hip down over the foot and up 2 or 3 in above the toes (Figs. 93 and 94). While the cast is being put on, it is important to keep the leg in



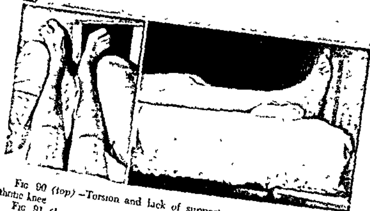


FIG 90 (*top*) —Torsion and lack of support increase strain on the arthritic knee

FIG 91 (*bottom left*) —Correct support and alinement obtained by sandbags and footboard provide maximal relaxation

FIG 92 (*bottom right*) —Early minimal contraction is prevented by proper placement of a folded towel

maximal *extension*, holding the foot in correct alinement and at right angles to the leg. A quick-setting plaster is used, and as soon as the plaster is set a top strip is cut from the cast with a *sharp knife*. *The* cast is then spread, making a

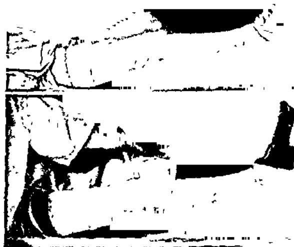


FIG 93 *(top)* —Rest cast for the arthritic knee. Sheet wadding adds to *comfort* *The* leg is held in maximal comfortable correction while the cast is applied

FIG. 94 *(bottom)* —Plaster is rolled on to the desired thickness, six rolls of 6 in plaster bandage being the average requirement (Posterior plaster *strips help* strengthen the cast.)

shell with high side walls (Figs. 95 and 96). (The high side walls are important to keep the leg from rolling out when *the patient* uses the cast for resting. The high foot construction is helpful in keeping the weight of the bed clothes off





FIG 95 *(top)*.—The cast is easily cut and spread open after the plaster sets and before it hardens or is entirely dry

FIG 96 *(bottom)*.—When the cast has dried it is lined with stockinet, sheet wadding is held in place by short strips of adhesive tape

the feet.) The cast is then removed, allowed to dry and later is lined with stockinet (Fig. 96). The cast is then ready for the patient to use as a rest cast in bed for as many hours a day as he can tolerate it comfortably. By complete support of the leg in such a cast, the patient gets maximal relaxation of the flexor muscles, and inflammation of the knee will subside most readily with this type of support. Periodically the patient is taken out of the shells to sit up on the side of the bed or in a chair for relaxation and for the exercises described later (pp. 145 ff.).

With an acutely inflamed joint one may choose to leave the leg in the solid cast (the strip is cut, but tied in, Fig. 97) for five or six days. This gives complete immobilization and will often help quiet an acutely inflamed or hot joint. On occasion the joint may be kept immobilized longer—up to one or two weeks, but not longer because ankylosis sets in rapidly. Although ankylosis will stop pain and is recommended by some, we do not advise it as long as the patient has a chance of maintaining a functioning useful joint.

When the patient has limited knee involvement but his knees are straight he is allowed to walk. If he cannot walk without limping, use of crutches is advised to relieve strain to the knee joints. A cane is never used (Figs 98 and 99). Crutches should have rubber pads and tips and must fit. To fit them, with the patient in good posture and shoulders relaxed, measure from the axilla to the floor at a point 8 or 10 in out from the foot. When using crutches, the patient does not lean on them but simply uses them for balance, applying the alternate crutch-to-leg method to keep a normal gait without limp (Figs. 100 and 101).

Even though the knees are strong enough for the patient

to walk, there may be enough active arthritis to be aggravated by climbing steps or sitting down on and rising from chairs; consequently steps and low chairs are to be avoided In such a case it is very helpful to elevate the toilet seat, (Figs 102 and 103) and put blocks under the bed and chairs (Figs. 104 and 105). With elevated seats and bed the pa-



FIG 97 —*When complete immobilization is required for a short period, the cast may be cut and the strip tied back in place, permitting easy conversion to a rest cast later*

tient can get up and down readily with a minimum of strain and damage to the knees. Low seats not only cause more strain when the patient gets up but promote bad posture and deformity (Fig. 105).

If the knee is swollen or somewhat unstable but it is decided that weight-bearing is not harmful, the use of a 3 in elastic bandage to give some pressure and additional support (Fig. 106) is often helpful This bandage must be



Fig 98 *(left)* —A cane gives no support to the knee and does not prevent limping

Fig 99 *(center left)* —Crutches help maintain good posture and normal gait by balancing.

Figs. 100 and 101 *(center right and right)* —For correct use, swing the crutches as one does the arms in normal walking The right crutch balances for the left leg, the left crutch, for the right leg The crutch parallels the foot. Avoid reaching out too far and taking too long steps

applied with care so as not to place it too tightly above or below the knee but yet give good firm pressure as it is drawn over the condyles laterally and medially

Hips—Involvement of the hips in rheumatoid arthritis presents one of the most difficult of problems Here again weight-bearing must be limited or at times forbidden Weight-

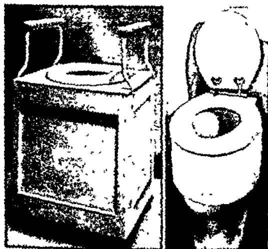


FIG 102 (*left*)—Commode built on casters to bed height. The back is open to permit rolling over the toilet
FIG 103 (*right*).—Wooden frame, covered inside and out with tin or aluminum, elevates the toilet seat to convenient height

bearing on inflamed hips adds strain to the joints which in turn increases muscle spasm and produces limitation of motion and progressive and increased loss of cartilage

With hip involvement it is essential to use a flat bed with boards under the mattress to keep maximal extension of the hips Periodically the hips are rested in different posi-

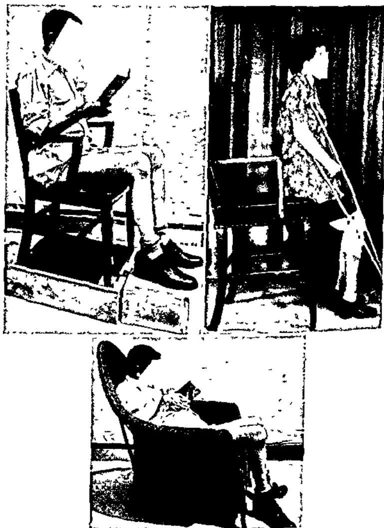


FIG 104 *(top)* —*Left*, box frame built to the desired height to fit a well selected chair The patient kicks the footrest aside when ready to stand *Right*, specially built chair designed to provide suitable height, firm seat and straight back and arms

FIG 105 *(bottom)* —Low chair means bad posture and joint strain when the patient gets up

tions—flexed, abducted, extended and rotated. The ... omes sits up periodically for maintenance of flexion of ... ndant and relaxation from rest in the extended position. ... pulling inflammation has subsided sufficiently and the ... of the allowed to walk, he must use crutches if he is ...

... y twist- ... found in ... ved joints ... strain of ... should be

... extremity is ... Character- ... side, elbows ... wrists flexed. ... the most pa- ... contractions ... the patient to ... with the arms ... arms so that

... possible in ... shed by plac- ... forearm and ... comfortably. The ... in an extended ... and



Fig. 106.—A properly applied elastic band

maintain correct posture and ... Crutches are used for balance ... on the hips.

Upper extremities.—... tremity is usually easier the ... is important to teach the ...

his body from the chair or the bed, as so many do when they have knee and hip involvement. When getting up from a low chair the patient is likely to raise himself by pushing up with his arms (Fig. 107). The fingers are flexed underneath



FIG 107 *(left)*—When rising from a low chair, pushing up with the ... extremity ... iw ... it, ... nt

to a standing position

the hand and are strained. The wrist and elbow also suffer a considerable strain in this maneuver. High chairs, high toilet seats and high beds help avoid this strain on the upper extremity Whenever it is practical the patient should ask for help in getting up from low places in order to reduce the strain A safe and easy method for the attendant and the

patient is shown in Figure 108. Note that the patient comes up with stiff legs to avoid strain on knees The attendant lifts by using his legs, knees bent, back stiff and pulling backward, letting his own weight do a good part of the work.

Many occupations that require lifting and heavy twisting of the arm or wrist (and many of these are found in household work) frequently add strain to the involved joints with inevitable damage. Occupations that require strain of the joints of the upper extremities obviously should be avoided

The greatest harm that comes to the upper extremity is usually from improper rest positions (Fig 109) Characteristically, patients are found with the arms at the side, elbows flexed, hands over the chest or abdomen and wrists flexed This is the most comfortable position and the one most patients seek, but it inevitably increases flexion contractions

When the shoulder is involved encourage the patient to rest periodically and as often as is convenient with the arms abducted For sitting, arrange a chair with high arms so that he may rest with the shoulders in abduction

The elbows too should be rested as often as possible in maximal extension On the bed this is accomplished by placing a small pillow or a folded towel under the forearm and extending the arm as far as it can be done comfortably The palm is turned up so that the wrist may rest in an extended position over the edge of the towel (Fig 110) A small pad or towel is placed under the upper arm to take pressure off the elbow In very acute stages of inflammation it may be helpful to make a plaster shell for the patient to rest in in maximal extension. Care should be taken to prevent loss of



FIG. 109 *(top)*.—Flexion contractions of the upper extremity are worsened by this position

FIG 110 *(bottom)*.—Folded towels are used, with thickness adjusted to give maximal comfortable extension

flexion of the elbow, for a fixed straight elbow is obviously worthless

Wrists notoriously develop wristdrop with flexion contraction A wrist that is in flexion definitely limits the patient's ability to make a good grip, whereas with a wrist that becomes ankylosed in extension the grip need not be limited (Fig 111). For this reason, with wrist involvement and a tendency to flexion contraction it is important to supply protection by means of a splint when the patient is up and about or for resting in bed when he is unable to rest in extension as already described The best splint for the wrist is a light weight cock-up made of a few layers of plaster The plaster can be molded well to the patient's arm, wrist and hand and can be extended up over the ulnar side to prevent the common ulnar deviation (Fig 112) The splint is best carried down to include support of the proximal phalanx By doing this we prevent excessive abnormal contraction at the metacarpophalangeal and proximal interphalangeal joints. The proximal interphalangeal joints that tend to hyperextend can be flexed over the end of the cast If the cast is made light weight and small enough, the patient can still use his fingers This is a great advantage, allowing the patient to wear the cast and continue many of his daily activities as well as pull the bedclothes over him at night

Spine —Although the spine rarely offers much of a problem in peripheral rheumatoid arthritis, attention should be given to good posture Standing too long, working in bad positions and sitting on low chairs with curvature of the spine increase strain and deformity To maintain correct posture and obtain the proper kind of rest, a flat bed is prescribed If the bed is not firm, boards should be placed full



Fig 111 *(top)*.—Wristdrop Grip is always poor.
Fig 112 *(bottom)*.—Plaster cock-up provides extension with comfort High side wall prevents ulnar deviation

length under the mattress. When the patient is allowed to sit up, a good straight-backed chair should be furnished that gives proper support to the spine Sitting for long hours at a time should be discouraged. Occupations that require the patient to bend over a table or a desk and such activities as making beds, ironing and leaning over a sink may increase fatigue and strain to the back

2. Proper Rest

Before prescribing the proper rest for a patient with rheumatoid arthritis, it is important to take sufficient time to review with him the different causes of strain and to outline a specific program and methods of avoiding them The amount and kind of rest, including splinting, will of course depend on the activity of the disease and the judgment of the physician Excessive or prolonged rest should be avoided because this leads to more stiffening and loss of motion Frequent turning in bed or change of position helps rest different muscle groups Sound sleep under sedatives aggravates the morning stiffness Aspirin, to promote relaxation and rest, is usually beneficial and sufficient However, aspirin should not be used to cover pain and thereby allow the patient to sit up longer or go beyond his physical capacity. Patients must be warned not to sit in one position oo long, an hour usually being the limit At certain times a patient may need five or six short rest periods during the day. As he improves, he spends less time resting and more time exercising. Regularity and balance of the daily schedule for rest and exercise must be followed

3. Exercises

Exercise is essential to the maintenance of function. In the presence of atrophy and the tendency to contraction that characterize rheumatoid arthritis, the prescription of the kind of exercise and the amount is of paramount importance. Unfortunately, too many patients are simply told to exercise as much as they can to maintain motion and strength without being given specific directions. It is a mistake to hand the patient a list of exercises and expect him to follow them properly without some instruction and follow-up supervision. The time taken at the beginning not only in teaching patients a good system of exercise but in explaining to them the reasons for exercise is well spent.

Reasons for exercise.—With careful explanation, any patient can be made to understand why the assigned exercises are important to his future well-being.

1. To maintain and increase motion Maximal range of motion will be maintained in joints only by putting every joint through its maximal range daily and up to tolerance. This helps prevent deformity.

2 To strengthen muscles that support joints. Exercises will build muscle strength Without muscle support, joints hang on their ligaments, causing chronic irritation, subluxation and deformity. Muscle support prevents deformity.

3. To increase circulation. Circulation in rheumatoid arthritis is notoriously bad, and regular exercises will do more to improve blood flow than any other treatment.

A patient with much involvement of the weight-bearing joints cannot walk far enough to preserve his muscle tone and strength In addition, the walking does not require full

range of motion of the legs. In bed, however, with daily use of a good set of corrective exercises without the irritation of weight-bearing, muscular development is unlimited. By using corrective exercises of all muscle groups he has a better chance of maintaining a full range of motion. Many patients say they get all the exercise they need through their housework or other occupations. Here it should be explained that most housework and many occupations produce strain on joints and may do more harm than good, and certainly do not provide for the full range of motion of each joint daily. If the patient is able and required to work he is much better equipped for work if he has developed good musculature through regular daily exercises.

Tolerance for exercise.—The amount of exercise prescribed is very important. Too often the patient says "Yes, I was given a set of exercises, but when they hurt my joints I quit." Here one must immediately explain to him that there is a definite amount of exercise every patient can tolerate without aggravating his joints. Because of atrophy and weakened muscles, the tolerance for exercise in a patient with rheumatoid arthritis may be exceedingly limited. It therefore is important that he follow a definite schedule of so many exercises per day on a definite training program. When the process is very acute or active the tolerance may be only two or three contractions of the muscle group before fatigue sets in. Practically every patient can start with two or three contractions of each muscle group without having trouble. This series is repeated two or three or four times daily, and after two or three days, if the patient is tolerating this amount, the number is increased by one. When this is tolerated, after three or four days, the number of exercises

is increased again. A 10 per cent increase at a time is safe. Exercising only once a day with three or four contractions is not going to build muscle or be sufficient to do any good, so the patient must be impressed by the fact that he must have at least two or more exercise periods per day to make any substantial gain.

RULES OF TOLERANCE.—1. Some muscle pain and soreness may be expected when the exercises are first started, but this will usually subside within a few days to a week

2. Pain during exercise is not significant.

3. Pain that persists for hours after the exercise period or increased pain with the same exercises the next day means that the exercises should be decreased until the added pain is gone The exercises are decreased, *not stopped* As soon as the increased pain has subsided the exercise again may gradually be increased as scheduled.

Tolerance for exercise varies, of course, according to the weakness and length of non-use of the muscles and the amount of inflammation of different joints Certain exercises can be pushed along much faster and in greater numbers than others, for example, exercises 1, 2, 8, 11, 12, 14 and 16 in the following list. The patient learns this as he progresses. As the patient gains strength, he is constantly encouraged to do more things for himself as long as the extra activities do not produce joint strain or unusual fatigue.

The prescription of exercise.—This must, of course, be individualized in each case, depending on the extent of the arthritis, the degree of activity, the general state of nutrition, strength, etc. A physical therapist trained in muscle re-education work will be able to carry on with the patient after simple directions from the doctor. Without the help

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of a physical therapist the doctor will be obliged to take more time to explain the list of exercises to his patient and to teach him at the start It is well to remember that deformity comes mostly in flexion, therefore extension exercises are stressed For simplification, active exercises that would be suitable for a patient with rheumatoid arthritis and multiple joint involvement are outlined as follows

Active Exercises for Rheumatoid Arthritis

1 Lying on back, legs straight Take a deep breath, expanding the chest. Hold the chest expanded until the count of 5, then relax.

2. Lying on back, legs straight. Contract the abdominal muscles, flattening the abdomen Hold the abdomen flat while breathing naturally.

3 Hands·

a) Make a fist

b) Stretch the fingers straight

c) Spread the fingers apart

d) Pull the thumb over to the tip of each finger, making as round a circle as possible

4 Hand resting on the bed, palm down, or on body if elbow will not straighten Raise the hand for wrist motion

5 Elbow bent at a right angle, upper arm resting on the bed

a) Bring the fingers to the tip of the shoulder.

b) Keeping the palm turned up, push the hand down ward the bed, straightening the elbow

6 Arms resting at sides, palms toward body

a) Raise the arm sideways away from the body, then back

b) Raise the arm, thumb leading, forward, upward and as back as it will go, then down to the side in reverse order

c) Lying on back, legs straight, arms at side Raise the arm ard, upward and as far back as it will go Then swing the ut to the side and around back to the side

Lying on back, legs straight Curl the toes down, keep the

toes curled and turn the foot in and pull the foot up, stretching the heel cord, hold the three motions until the count of 5. Then relax the whole foot.

8. Lying on back, legs straight: Contract the muscles on top of the thigh, pulling on the patella (kneecap) and flattening the knee. This exercise is increased up to 10 times a day, 10 times each, if knees are particularly involved.

9. Lying on back, legs straight: Holding the leg straight, take the leg 15 in. to the side and back. Do not go far enough to tilt the pelvis.

10 Lying on back, knees bent, feet on the bed: Raise the knee toward the chest, straighten the knee by lifting the foot in the air, stretching up with the heel; let the knee bend and the foot return to the starting position. Alternate knees

11 Lying on back, knees bent, feet on the bed: Pull in the abdomen, then squeeze the buttocks muscles as if to roll the seat up off the bed, tilting the pelvis and flattening the lower back hard on the bed.

12. Lying on back, legs straight. Flatten the neck by making a double chin, and at the same time stretch up through the top of the head as if to pull the ears away from the shoulders.

13 Lying on back, legs straight: Turn the cheek as far as possible toward the bed, stretch, keeping shoulders flat on the bed, come back to straight position and rest.

14 Lying on back, legs straight: Pinch the shoulder blades together.

15. Lying on back, legs straight, arms at sides with elbows and backs of hands on the bed. Keeping the hands and elbows as near the bed as possible, push the arms up until the upper arms are beside the ears, stretch; pull down to the sides in the same manner.

16. Lying on back, knees bent, feet on the bed: Take a deep breath, expanding the chest, hold the chest expanded and push the diaphragm up and down five times (panting).

17. When the knee has a severe flexion deformity, add knee

extension in the sitting position on the side of the bed (contracting muscles on top of the thigh and pulling the patella)

Again it should be noted that these exercises can be varied according to the degree and extent of the involvement in each patient. Some patients will not need or be required to do all of these exercises, others may be given additional exercises to suit their particular needs. In any event the patient should be furnished with a typewritten list of the exercises he is to do, with specific instructions how to do them and how many to do daily He should be given specific instructions about reactions and rules of tolerance so that he does not stop because the joints begin to hurt

After the patient is given the first course of instruction and a list of the exercises, he practices the exercises for a week and then is observed to make sure he is doing them correctly. It may be necessary to continue checking some patients weekly or perhaps only monthly to make certain that they are doing their exercises up to their tolerance It is well lso to inquire into the amount of time spent at exercises ecause many patients tend to hurry through them The erage patient builds up to two or three hours or more of ercise daily.

The patient, of course, should be instructed to establish egular schedule for his exercises, doing them at regular rs each day All exercises are done slowly, with complete xation between each exercise As the patient improves, tance can be added

he patient who has not had regular exercises and who ad the disease for any length of time will usually be sed and much pleased to note how much he has ac-shed in the first month or two on a regular exercise

tient is encouraged to use more of his own muscles, making a combined active-passive exercise. With the development of still more tolerance the therapist can give extra push or traction to gain maximal motion up to normal range.

ANKYLOSED JOINTS

When there is no joint motion, exercise is necessary to maintain the best state of nutrition and prevent the trophic disturbances and dependent edema that come with inactivity. This type of exercise is called muscle tensing and is accomplished by the patient's learning to contract or set his flexors against his extensors By the tension muscle exercises the patient not only maintains a much better state of nutrition than he otherwise would have but is usually much more comfortable, with less soreness and aching of muscles Exercises 1, 2, 8, 11, 12, 14 and 16 (pp. 145 f.) are important for all patients with rheumatic disease but are specially mentioned here because they can be done in spite of joint ankylosis.

Pool exercises.—Exercises under water are, as a rule, highly beneficial and their practice is encouraged whenever possible. In patients with subacute arthritis and in the presence of much muscle tension and spasm, water promotes relaxation Exercise under water is especially good when the weight-bearing joints are involved The patient gets much better knee and hip motion than he could in bed or from walking and has no strain of weight-bearing. The Hubbard tank is useful, but a pool shoulder-deep affords better chance for walking and maximal extension. The patient should be

cautioned firmly against (1) overdoing, and (2) chilling.

Relaxation exercises

1. Take a deep breath, fully expanding the chest, hold the chest expanded until the count of 5, then let the breath out and relax completely, breathing naturally
2. Stretch the right arm down, stretching, stretching, to the count of 1-2-3, then relax the arm completely and make the entire body become relaxed and limp
3. Repeat with the left arm
4. Stretch the right leg down, stretching, stretching, then relax the leg and whole body
5. Repeat with the left leg
6. Stretch the neck, flattening and stretching right up through the top of the head, then relax the neck and whole body
7. Rest 20-30 seconds after each stretching exercise, and repeat the list two or three times

Occupational therapy, aside from its diversional and possible economic rehabilitation aspects, can be a highly useful adjunct in the preventive and corrective exercise program

General physical activities and working should be encouraged but guided in the kind and amount by the rules of tolerance. Work or outside activities should be undertaken at *regular* hours, with a gradual increase to as much as the patient can do without undue fatigue and strain of joints

During a flare-up, patients tend to do nothing for weeks or months, then, when feeling better, suddenly do much more than they are conditioned for and consequently strain their joints and induce another relapse

A safe rule for increasing physical activity is to add 10 or 15 per cent to the total for the day, and be sure of tolerating this for several days before adding another 10 per cent

SPONDYLITIS

The deformity in spondylitis is nearly always characterized by a flexion of the spine, rib cage contraction and loss of normal posture with rigidity. The spine in spondylitis will ultimately ankylose in varying degrees, the extent depending on the stage at which the disease is arrested. Ankylosis usually starts in the lumbar segment and spreads upward, with early involvement of the dorsal spine and loss of chest motion Frequently it affects the entire spine, including the neck The serious crippling comes from a frozen flexed spine, the patient drawn over with face to the ground, unable to sit or stand erect.

A straight, stiff "poker" spine is not a serious deformity. After the active disease has subsided the stiff straight back does not interfere with the execution of most jobs and normal living

The acute form may develop rapidly with so much muscle spasm and pain that immobilization in bed or application of a cast or both is necessary, along with other appropriate measures (p. 56). Rest should, of course, be on a flat bed to maintain maximal extension (Fig. 89, p. 125). A posterior rest shell (body cast), carefully made with the back as straight as possible, is highly satisfactory for resting and relaxing the spastic erector spinae muscles.

Hip and shoulder involvement, followed by severe limitation of motion, is frequently seen in the more severe types. In a few instances the process finally invades peripheral joints, causing the typical changes of rheumatoid arthritis.

The chronic form, with insidious onset and progress, is much more common. This type, not too disabling at the

onset and much slower to cause deformity over a period of years, requires much more understanding and co-operation of the patient. The first step in prevention of deformity is a thorough explanation of the disease, so that the patient years hence does not discover too late that he has a deformity that cannot be corrected By faithful adherence to the correct preventive program, the back can be kept straight Rest to keep normal posture is prescribed to fit the stage of the disease. A flat, nonsag bed, with boards under the mattress, is essential for all

The amount of rest depends on how fast the patient loses normal posture and develops fatigue In the more or less acute or subacute stages the patient may not be up more than an hour before he begins to "droop" He is advised then to rest flat on his back for 10–15 minutes, or as long as necessary to regain normal posture, before getting up again During the time he is up, the patient should use straight-backed chairs, and occupations that require poor posture, as leaning over a desk, bench or the like, are to be avoided Patients who work away from home can often arrange to have a cot available for short posture rests during the day, and are thus able to stay on the job When this is not practical and the patient must work beyond his tolerance, a Taylor brace or body cast is prescribed However, when possible, the rest program is better In ambulatory patients, a good method for watching posture is to have the patient stand back to the wall, heels and buttocks touching, and then measure the distance between head and wall Chest expansion, measured with a tape, can also be quickly recorded

Hip and shoulder involvement may come on insidiously and cause rather severe limitations When these joints are

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affected, preventive measures should be started at once, as outlined for rheumatoid arthritis.

Exercises.—In spondylitis, corrective exercise is not as effective in maintaining motion of the spine as is exercise in the peripheral joints Every patient, however, should take corrective exercises daily to preserve normal posture, chest expansion and hip and shoulder motion (see the list of active exercises on pp. 145 f.). Special emphasis is given to exercises 1 and 2 and 7–16. One of the most important is chest expansion. The same rules of tolerance apply for spondylitis as for rheumatoid arthritis.

DEGENERATIVE JOINT DISEASE (OSTEOARTHRITIS)

Prevention of deformity in this type of arthritis is important, but not nearly as much of a problem as in rheumatoid arthritis Osteoarthritic deformities are most likely to appear in the hips, back, hands and sometimes the knees Hip involvement, malum coxae senilis, is the most common and at times the most serious problem In the acute or subacute form with much pain and limitation of motion, weight-bearing and walking must be limited or even forbidden for a while The use of crutches is nearly always prescribed to relieve weight-bearing in the affected joints. With crutches the patient can learn to put the leg through a walking range to preserve motion and strength without joint irritation. Canes are not satisfactory. The principles and advantages of protecting the joint, saving what cartilage is present and maintaining motion for the rest of the patient's life, must carefully be explained. Many are grateful for the knowledge that they can do much more with crutches and with less discomfort than without them.

Frequent periodic rest on a flat bed is prescribed to maintain extension, the amount of rest varying with the stage of the disease and age, strength, etc , of the patient

Corrective exercises are just as important as in rheumatoid arthritis and the same rules apply (pp 145 f.) Assistive stretching to keep extremes of range of motion is sometimes helpful Surgery is rarely, if ever, indicated

Spinal involvement is extremely common, but disability is minimal Patients should be warned about the dangers of trauma, hard labor and lifting, especially lifting with rotation Patients are taught to set their backs rigid and lift objects by bracing their backs (bending hips and knees). Gardening, hoeing, weeding, etc , often cause trouble Firm beds and boards under the mattress, if necessary, are prescribed Straight-backed chairs are best, and patients are advised not to sit too long in one position (automobile riding included).

Corrective exercises to keep the back strong is good insurance against strain or trauma of ordinary activities

The joints of the fingers and hands are often involved but seldom become seriously crippled, although at times motion is quite limited. Here the usual rules apply regarding avoidance of chilling, strain in occupation, etc Keeping warm, wearing gloves, using corrective exercises are some of the measures always prescribed

BURSITIS

Bursitis is common in many locations, but only in the shoulders is it very disabling Bursitis is often mistaken for arthritis because in the shoulders, like arthritis, it can be acute, with pain and pronounced limitation of motion, or

develop gradually with a slow progressive loss of function Unchecked, it can go on to ankylosis of the joint and severe atrophy. Early diagnosis of bursitis is important to prevent loss of motion.

Start rest periods in abduction and external rotation. Add corrective exercises and include "wall-creeping" daily to maintain motion *Wall-creeping is best carried out by standing* facing a wall with the toes about 12 in out from the wall. The hands are placed on the wall as high as possible and alternately, using the fingers, creep up the wall as far as the patient can reach. The exercise may be done with one hand at a time, but simultaneous use of the two hands is preferable because it achieves better balance. By early establishment of the routine, nearly all disability can be prevented Shoulder motion that is already limited usually can be worked out by assistive, corrective exercises and stretching carried out by the physical therapist. The application of heat to the shoulder—hot compresses, heat lamp, electric pad or diathermy—for a half hour before the daily stretching will often prove helpful.

Shoulders in which motion has been limited for a long time are more resistant to daily physical therapy, and manipulation under anesthesia may be necessary to break the adhesions. It must be remembered that daily passive and assistive exercise must follow manipulation in order to maintain motion.

CORRECTION OF DEFORMITY

Unfortunately, patients in a search for relief from pain rest in flexion and develop contractions before they or the doctor realizes what is happening In the beginning, the

slight contraction may go unnoticed, and further contraction is so insidious that it may be missed until it becomes pronounced. Frequent checking by the physician, with the patient stripped so that his range of motion is clearly observable will help detect early limitations At this time, corrective measures are relatively simple, i e, proper rest, extension and exercises to gain motion.

For early contractions of only a few weeks' or months' duration, the simple measures may be all that is necessary

Do not wait until active arthritis or inflammation subsides to start corrective procedures If the joint is acutely inflamed it may be necessary to wait a few days or more, but with delay until all signs of activity and sedimentation rate have subsided, it will probably be too late Some activity may persist in joints for years

More severe contractions and those of longer duration require more strict attention The basic principles of rest and relaxation are followed Gentle, firm, daily assistive exercises and stretching are helpful, but care must be taken not to irritate the joint by use of too much force, secondary irritation and pain will cause more contraction and "splinting" of the joint For this reason, we object to the use of traction or wedge casts To obtain lengthening of the flexors, the flexors must be comfortable They must relax to gain maximal extension

SHOULDERS

The shoulders are rested in abduction and extension as frequently as indicated and tolerated Assistive exercises and stretching are practiced daily, use of a heat lamp, heat pad or hot packs on the joints before extension and stretching

may help. The degrees of range of motion are measured every two to four weeks. As long *as improvement in range* is noted on routine check-up, the procedure is continued. When and if no further progress can be made and it is still necessary to gain more motion, manipulation under anesthesia should be considered. This procedure, however, requires training and experience on the part of the operator and skilled physical therapy help afterward. Shoulders are usually very painful after manipulation, and even a well trained physical therapist is not always able to maintain increased range of motion in this joint. Success also depends on choice of the optimal time for manipulation and the stamina of the *patient to go through with it*

ELBOWS

Elbows present a real problem. They are especially difficult to keep mobilized after manipulation. If there is ankylosis or serious limitation, surgical resection by an experienced orthopedist may be considered.

WRISTS

Deformed wrists are nearly always in flexion, wristdrop, and sometimes in ulnar deviation. Nothing need be done about an ankylosed wrist that is straight or in extension, as these positions are optimal for the use of hands and fingers, making a grip, etc When a wrist is flexed sufficiently to impair the use *of the fingers in grasping objects and in* other necessary activities, but is not too firmly ankylosed, it can be reset in a more useful extended position under anesthesia.

HIPS

Hips that are contracted or have serious limitation of motion are also difficult to manage. They respond poorly to force, especially if there is joint irritation because of weight-bearing or active arthritis Fortunately, hip involvement is not as common as deformities and limitation in other joints To gain hip motion, weight-bearing must be curtailed or forbidden altogether until signs of joint irritation, muscle spasm and pain subside. With a minimum of joint irritation, more force can be used in assistive stretching along with bed exercises Exercise and stretching under water are especially beneficial.

If after the signs of acute activity of the arthritis have subsided, assistive stretching and bed exercises have not given enough motion to permit walking or sitting, or both, manipulation under anesthesia should be considered. This procedure has definite limitations in that it should be done only by an experienced physician and when the requisite physical therapy facilities are available for proper follow-up. To mobilize the hip joint, arthroplasty may be indicated in a limited number of patients in whom the foregoing procedures have failed This operation has not proved too successful, giving only a limited range of motion in approximately 50 per cent of cases

KNEES

Knee contractions cause more serious disability in more people than any other deformity attributable to arthritis With methods now developed this situation should in the

future be reversed. First, all knee contractions could be prevented if they were treated early by the methods outlined earlier in this chapter, under prevention. Second, a method for correction of knee contractions has been devised that gives good results.

Routine for contracted knee.—1. Stop weight-bearing. Every step on a bent knee aggravates the condition and leads the patient that much faster to bed or wheelchair. Therefore, as long as there is a chance of correcting the contraction and restoring function, the patient should not be allowed to walk. Every contracted knee should have the benefit of an attempt at early correction unless there is obvious bony ankylosis.

2. Make a rest cast to hold the leg in maximal extension (Figs. 93–97), using the technic outlined on page 126. Have the patient rest in the cast as many hours in every 24 as possible. Casts must be comfortable to give maximal relaxation of the flexor muscles.

3. Start corrective exercises at once and insist on their being pushed to tolerance. Emphasize the quadriceps set, or "patella pull" (exercise 8, p. 146), having the patient do this exercise up to several hundred times daily.

4. As the flexors relax and the leg straightens, add pads *under the ankle* inside the cast to take up the slack and gain more extension.

5. If contraction at the start has been severe, a second or third cast may be necessary to fit the leg as it extends.

6. Measure the knee contraction every two to four weeks with an anglometer or simply the distance from the top of the patella to a hard table, to keep a record of progress (Figs. 113 and 114).



Figs 115 and 116 —Six years after manipulation, both knees straighten
ı 180 degrees and flex past 90 degrees in free motion without pain Despite
ınspicuous subluxation, cartilage loss and porosis, this youth, aged 21,
alks satisfactorily

7. Continue the foregoing routine until the leg straightens to 180 degrees or has reached maximal extension. If on the monthly check-up, measurement shows no further improvement and the knee is not straight, manipulation is indicated.

8. Manipulation is performed under anesthesia, only after the foregoing routine has been followed and then only by one trained and experienced in the technic. Proper physical therapy facilities for follow-up care are *essential*.

Manipulation.—The degree of damage of the knee visualized on x-ray examination is not a reliable guide in choosing cases for manipulation. Some of the worst cases, by x-ray criteria, have resulted in well functioning knees; others that did not look too bad have been the most difficult. From x-ray and other examinations it is impossible to predict the results that will be attained (Figs. 115 and 116). It can be emphasized, however, that the longer the duration of the contraction, the less chance there is of good results because of tighter muscles, adhesions and increased joint damage.

Although we cannot pretend here to teach the details of the procedure, a brief outline of our method is presented We insist first on carrying out steps 1–7, outlined above. After this preparation, which means two to three months at a minimum, the patient is hospitalized. Under general anesthesia the bent knees are manipulated to the best correction possible, usually they can be straightened to full 180 degrees Solid leg casts are applied, cut and tied (Figs. 93–97). After 24–48 hours, acute pain is gone and narcotics are no longer required Muscle tensing exercise is started at once (in the cast). After five or six days the leg feels loose in the cast. The cast is then opened, the leg removed for a few minutes, and

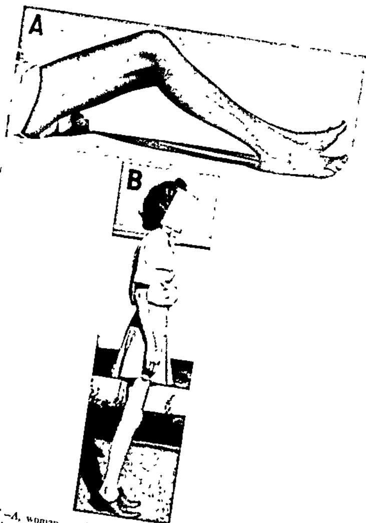


FIG 117 —*A*, woman, aged 31, with arthritis for seven years, has had knee contraction of 55 degrees for 18 months *B*, three years after manipulation, she walks with normal motion and gait

the physical therapist starts passive exercise through the greatest range of painless motion The leg is returned to the cast until the next day Each day the motion is increased to tolerance and active-assistive exercises are added Usually in four to eight weeks the patient is strong enough through graduated active exercises to hold his legs straight and flex the knees When sufficiently strong, he begins to stand without braces, but using crutches for balance. Careful daily supervision is necessary to teach proper posture, then joint motion, standing in place and finally the normal gait. With this program and follow-up, we have patients who are walking as long as 14 years after manipulation By maintaining good posture and following the rules of rest, exercise and avoidance of strain, there is no excuse for a recurrence of flexion contracture (Figs 117–119).

In 65 consecutive cases of manipulation on knees, reported previously, there were severe contractions and attempts at straightening by simple measures had failed. Some patients had undergone attempts by other methods elsewhere, without success. Age range of the group was 17–68 years Average duration of arthritis was 7.2 years and average duration of contraction 3.2 years. These patients were doomed to a wheelchair or bed. After manipulation, 40 walked normally, 12 walked with crutches unaided, 11 walked with crutches but required some help and 2 had ankylosed joints. Sixty-three of these patients were walking an average of eight years after manipulation.

Many of these patients had active arthritis in the knees at the time of manipulation, but we have never seen a flare-up or exacerbation produced by manipulation of the knees Frequently, it is noted that active arthritis in the joint sub-

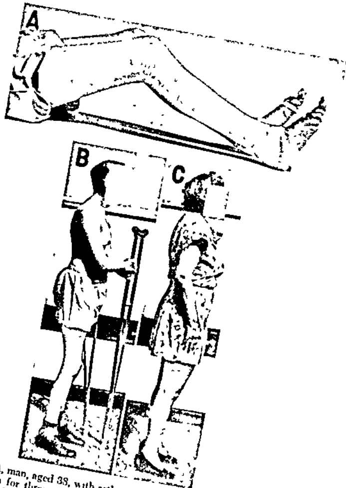


Fig. 119.—A, man, aged 38, with arthritis for 23 years, has had 55 degree nee contraction for three years. (The left hip is contracted, with 75 per nt loss of cartilage.) B, same patient as preceding, walking with normal tion of the knees 12 years after manipulation. He uses crutches to protect bad left hip and maintain motion in it. C, woman, aged 53, with arthritis nine years and 60 degree knee contraction for one year, now walks with nal range of motion seven years after manipulation.



Fig 118—*A*, woman, aged 32, with arthritis for eight years, for four years has had knee contraction of 50 degrees, with pronounced loss of cartilage and secondary hypertrophic changes *B*, two years after manipulation she walks with normal motion and gait

foot with relaxed arches that are sore and tender (especially metatarsals), so common in rheumatoid arthritis, needs correction when weight-bearing is permitted Often the large sponge rubber metatarsal pad, properly fitted and located in the shoe, will be sufficient When not, the metatarsal bar is prescribed (p 124). A proper-sized shoe, strong and large enough, and with a straight last to prevent crowding of the toes, can usually be found in various makes. The heel should be low or not more than medium height The longitudinal arch support, also made of sponge rubber, must be of the proper size and located in the shoe with care When the deformity is more severe, with eversion of the foot, wedging may be necessary This is usually started with a ⅛ in leather wedge inserted into the heel and sole by the shoemaker If ⅛ in wedging is tolerated, but not sufficient to give correct alinement, another ⅛ in. wedge is added

The stiff or rigid foot, cold, and often very painful, can be helped dramatically at times by manipulation, in addition to the exercises Two or three times a week the therapist or physician firmly grasps the tarsal bones (long arch) with one hand and gives one firm rotary twist of the metatarsals in each direction with the other hand With this maneuver one may hear the popping of many adhesions The patient should be warned that the first few times the maneuver may be quite painful momentarily, and he must relax the foot as much as possible for best results In a short time with this procedure the foot is often noticeably warmer and less painful When joints are acutely inflamed, manipulation is deferred.

Hammer toes, also extremely common, can often be cor-

sides after manipulation. This is probably due to removal of joint strain by placing the joints in a more normal anatomicophysiologic position and complete support for several days to a week in a cast.

Surgery.—Posterior capsulotomy may be resorted to in the badly damaged and subluxated knee. We have used surgery less and less as we have gained more experience with manipulation.

ANKLES

Ankle deformity is usually in the form of footdrop. Again, in the early cases much can be done by daily stretching of the Achilles tendon by the physical therapist. In long-standing cases with severe contractions, manipulation under anesthesia will often be sufficient to correct the shortening. When this fails, a tenotomy by the orthopedic surgeon may be indicated.

FEET

Deformity in feet is not only one of the most common developments but one of the most neglected. The wearing of poor shoes without support, or bedroom slippers, may lead to flattening of the arches and eversion of the foot. With tight shoes and no exercise, the rigid or boardlike foot develops, often with hammer toes, bunions and related deformities.

A foot that is only flat, but still strong, and has motion and good alinement for weight-bearing is usually of no great concern. If it is asymptomatic and function is good, correction should not be attempted with arch supports. The flat

Aids to the Cripple

Crutches —Handles on crutches can be angled, moved up or down or placed on the outside to facilitate easier grip by the deformed hand The lower the handle, the better for extending the flexed elbow. Rubber tips prevent slipping and rubber shoulder pads decrease shoulder soreness (See also the note on fit of crutches, p 130)

Wheelchairs —The type of wheelchair selected should fit the patient, to assure proper posture They are for moving the patient and are not good for long hours of sitting A sturdy kitchen chair with large casters is often useful about the house The quadriceps develop as the patient propels himself backward Commodes on casters built to bed height and with open back are often helpful in moving the patient to the toilet or for use beside the bed (Fig 102, p 133).

Ramps —The difficulty of negotiating a few steps can often be overcome by placing a simple wooden ramp

High chairs.—These help avoid much strain in getting up and down. Chairs with straight backs and arms are best and can easily be elevated to proper height by 6 × 6 in wood blocks Extrafirm cushions may be used to raise the seat A small box or footstool is then necessary to support the feet (Figs 104 and 105, p 134)

High toilet seats —Proper height is easily obtained by removing the seat, building a simple wood frame on top of the bowl and placing the seat on top (Fig 103, p 133)

Automobile riding —To get into an automobile, back up to the front seat, sit down and pull the legs in afterward

Bedside tables —Many types are available One with adustable height and tilt top and book rest is an advantage in eeping good posture during reading or writing Some are made turn upside down and have clamps to hold reading material r patients who must lie on their backs

Wrist cock-up —A simple splint can be made with 3 × 10 in ca board padded with bandage A 3 in roller bandage is

rected by early stretching of the extensors by the technician. The technic is simple. Grasp the left longitudinal arch with the left hand, placing the thumb under the distal head of the metatarsal, then take a firm grip on the corresponding distal phalanx, apply traction to separate the joint and pull downward to stretch the extensor tendon. One good firm pull is painful for a second but becomes less so on subsequent maneuvers if done two or three times weekly. The same procedure is applied to each toe requiring it, remembering to give adequate support and traction before flexing The hands are reversed for the opposite foot. Increased or persisting pain and soreness indicate either that too much force has been used or that the arthritis is still too active to justify the maneuver.

In long-standing contractions that cannot be relieved by the foregoing methods, surgery with resection of the distal metatarsal heads may be indicated. In selected cases, properly done, the resultant rehabilitation is quite gratifying

While we search for a cause and cure, prevent deformity.

Index

NOTE —An asterisk (*) following a page number indicates reference to an illustration

Index

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strapped on top of one end for cock-up and hand rest. It is applied with a short 2 in. elastic bandage.

Rubber shoelaces.—These are helpful, used in Oxfords, for patients with hip or knee involvement that prevents them from reaching their feet.

Handy stick.—Many variations are easily made. One on the market has a hook and ring on one end for picking up articles, pulling on socks and trousers; in the other end is a magnet which proves very useful.

Telephone.—Bedridden, ankylosed patients can have a handpiece mounted next to the head of the bed and a button switch placed at the finger-tip for control.

Electric blankets.—These are light weight and very helpful in keeping even temperature.

Cradle.—A large paper carton or wire frame takes the weight of bed clothes off sore joints.

Pillow splint.—A very hot or acutely inflamed joint can be temporarily splinted by folding a large pillow around the extremity and bandaging it in place.

Cellucotton.—This comes in sheets about 2 in. thick and can be cut to desired size and shape for relieving pressure sores. It is cheap and disposable.

Sponge rubber, foam rubber.—These are good for seat pads and are also available in various thicknesses and sizes for mattress pads.

With a little effort and ingenuity the home mechanic can make other aids and accessories to facilitate rehabilitation and relieve strain on the joints.

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